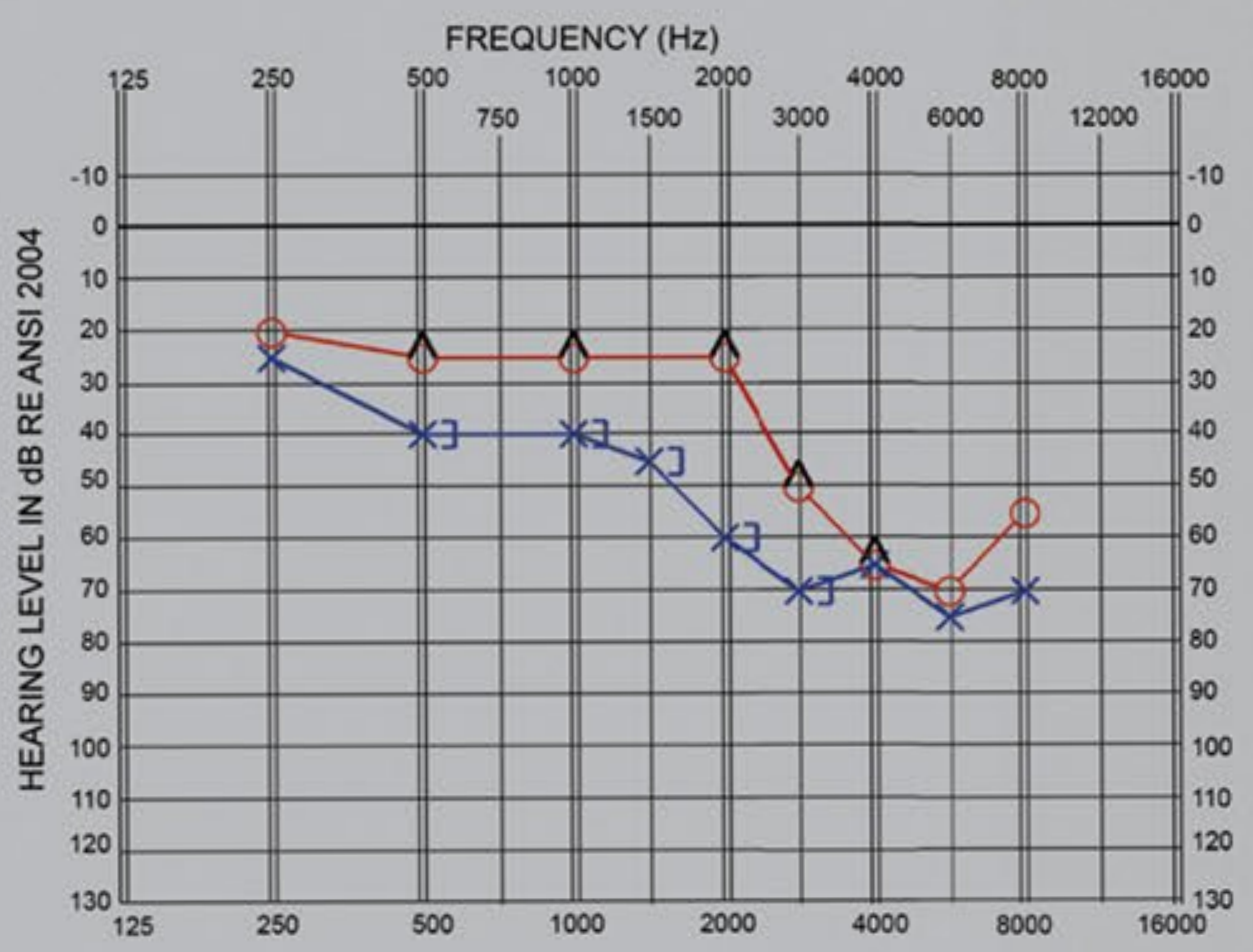
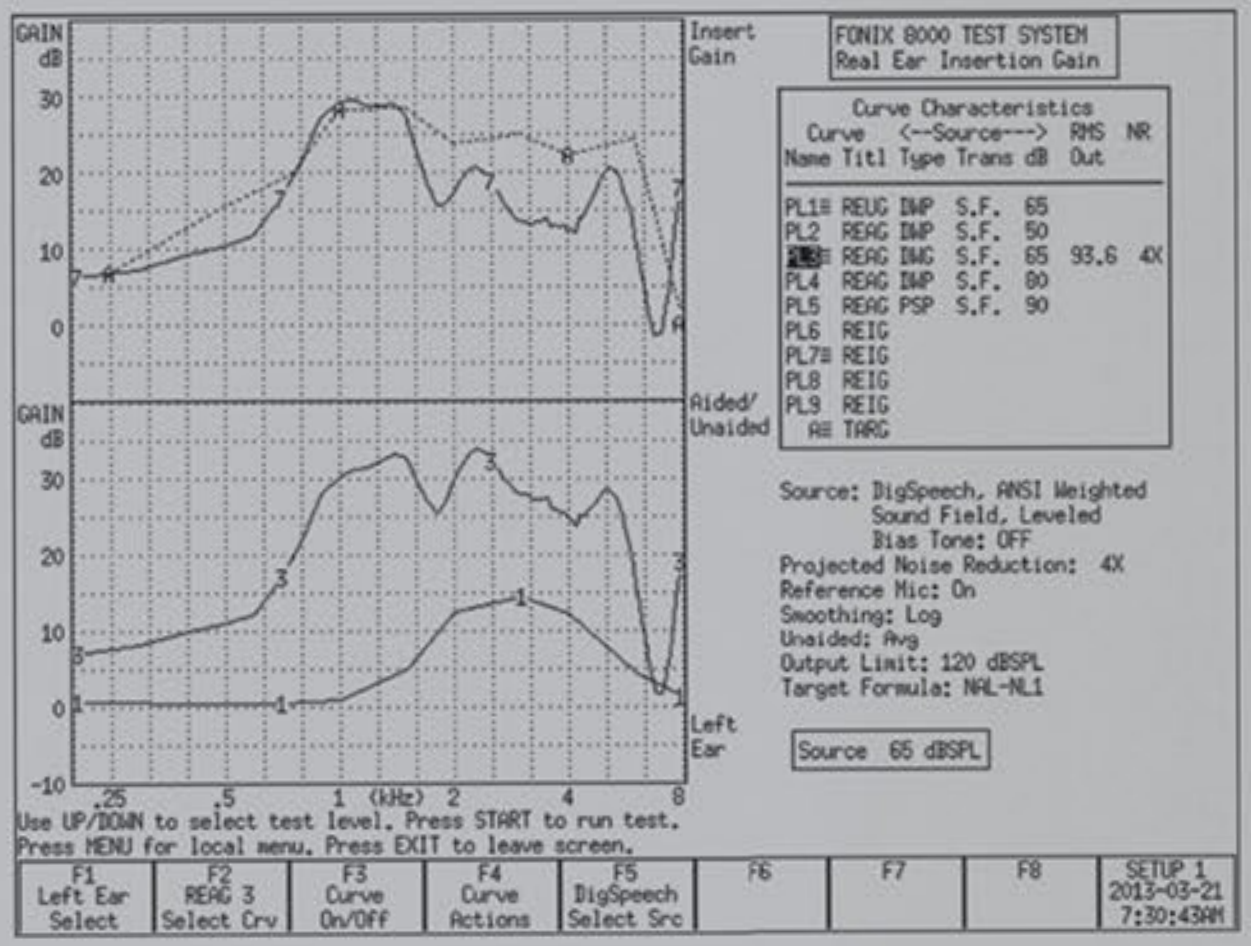
Adult Audiology Casebook

Michael Valente
L. Maureen Valente





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FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out

PL1≡ REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3≡ REAG DWG  S.F.  65  93.6  4X
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7≡ REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Avg
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

 Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.
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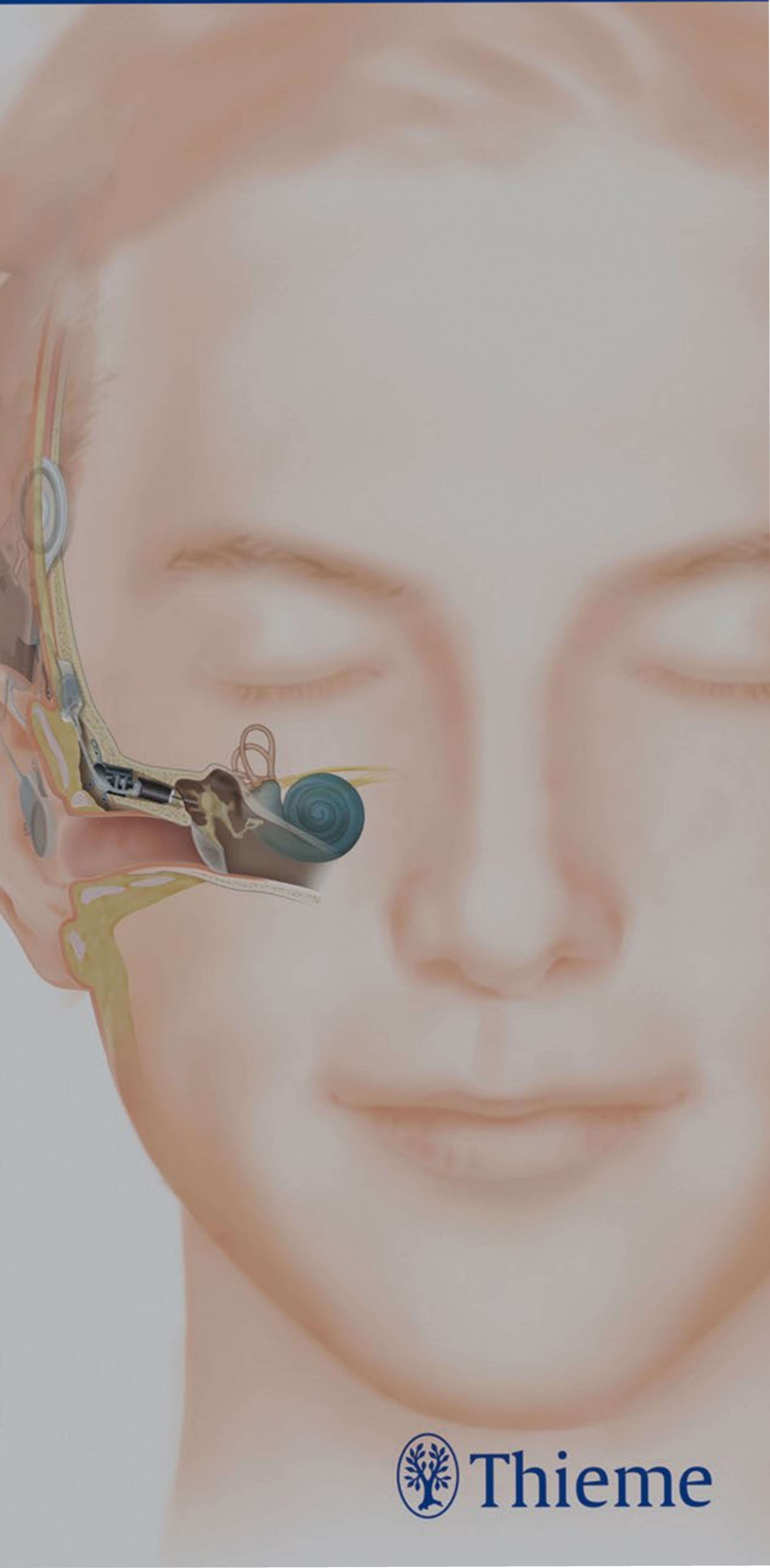

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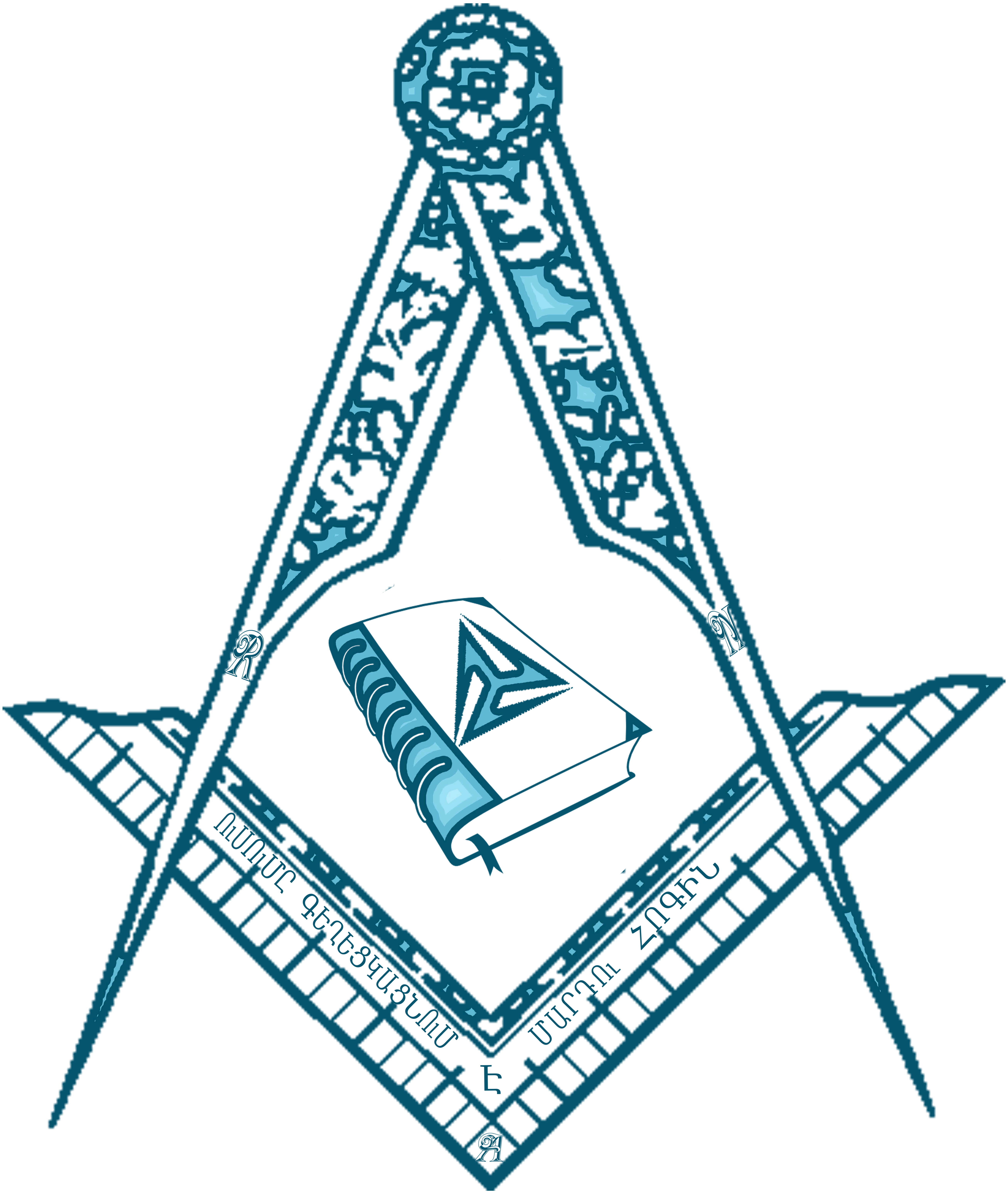
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Adult Audiology Casebook

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Preface

Several years ago the authors were approached by a representative from Thieme Medical Publishers to assess our interest in editing an adult version of the excellent textbook "Pediatric Audiology Casebook" by Jane Madell and Carol Flexer. At the time we were very busy with a number of projects and the responsibilities of our positions at Washington University that we informed the representative that we would have to give this some considerable thought. After giving the idea some consideration, the authors accepted the offer. Our first step was to obtain a copy of the Madell and Flexer textbook and review its contents to determine how these two editors organized each case. We decided to adopt the organization of the Madell and Flexer textbook with a few additional sections. We also decided that one of the most important elements to the reader was consistency in presentation across all the contributions.

At this point the authors decided to divide the cases into eight major categories. These categories included hearing disorders, diagnostic examination-auditory function, diagnostic examination-vestibular function, amplification-hearing devices, cochlear implants, hearing assistive technology, tinnitus management and other rehabilitative strategies. We then forwarded to colleagues around the globe a lengthy e-mail with information concerning the following: targeted audience of the text (students, clinicians and otolaryngology residents), an overall introduction to the textbook and its goals, desired divisions within each case report, and author guidelines for consistency across case reports. We also informed potential authors of our desire to create a new software application for each author to use so that the audiogram in each case report would be uniform. Finally, we communicated that the textbook would be exclusive to case reports of adult patients and outlined the eight general areas.

The e-mail was forwarded to numerous colleagues around the world, as described, and we were gratified at the initial response. This led to the securing of 68 authors from around the world and the 71 case reports you see enclosed between the covers of this textbook. Each case report was divided into sections involving (a) clinical history and description which is a one to two sentence introduction to the case report, (b) audiologic testing completed on the patient, (c) questions to the reader based on the introduction and the findings from the audiologic examination(s) described in the previous sections, (d) discussion by the author(s) of questions presented to the reader, (e) additional tests that may have been administered, leading to additional questions and resultant discussion, (f) description of the final diagnosis and treatment options that were either recommended or administered, (g) a brief description of the final outcome of the case, (h) summary of key points to be learned from the case, and finally (i) listing of a limited number of key references.

Each of the initial versions of the 71 case reports was reviewed three times by the editors. After the reviews were completed, our comments and suggestions for a revised version were forwarded via e-mail to the authors. When completing the review process, the editors wanted to ensure that our comments and suggestions were reasonable, to allow the next version to be more consistent with the other case reports, and to ensure that technical aspects would be beneficial to and appropriate for the intended audiences. We believe the case reports meet those goals.

As was mentioned above, to prepare for this textbook a new software application was developed by the authors and Todd Sproul, PhD, a software engineer at Washington University in St. Louis. Our goal was to be sure the audiograms in the case reports would be uniform and adhere to current standards. The audiogram software application was given to each author as a small token of our gratitude for each author's effort and dedication to the project.

Preparing a textbook is a two- to three year journey. The journey, like any other of life's journeys, was mixed with joy, pride, satisfaction and frustration. There were moments of triumph as well as moments of defeat. Some contributors elected not to submit the promised cases. Others submitted case(s), but the content was not in line with the goal of the textbook. Some decided not to submit the number of initially promised cases, while others, thankfully, submitted more than the original number of promised cases. The one commonality was that all authors reviewed the suggestions of the initial review and all submitted a revised version that did not require an additional version to be completed and submitted.

We are pleased with the final outcome and the diversity of cases, representing the depth and breadth of audiology's scope of practice. As the reader may see, some examples may be more routinely seen in the clinical setting, while other examples are highly unusual. Our goals are to educate and to help stimulate critical thinking and clinical insight development on the part of the reader. As we move forward with our doctoral profession and a greater degree of autonomy, may we continue to realize that each patient is different and an individual. There are no cookbook answers or solutions; rather, the critical thinking piece is crucial as we determine how to provide the utmost in high-quality care for our listeners with hearing loss. May these examples also provide guides in development of cases and professional writing style.

The editors would like to thank all 68 authors who contributed their time, expertise, and patience into preparing their case reports. Without their contributions, this textbook would not have been possible. We would like to give an additional thank you to Amyn Amlani, Michael Cevette, Susan Fulton, Lauren Harvey, Lisa Lucks Mendel, Heather Monroe, Kristi Oeding, Belinda Sinks, Brad Stach, and Wayne Wilson who agreed to our request to consider submitting additional case reports. These pleas arose as a result of a few contributors who for one reason or another were unable to submit the original number of agreed- upon case reports.

The editors would also like to thank Richard Chole, Chairman of the Department of Otolaryngology Head-Neck Surgery at Washington University in St. Louis School of Medicine and William Clark, Program Director in the Program of Audiology and Communication Sciences at Washington University in St. Louis School of Medicine for allowing the time and support required for the editors to complete this project. We would also like to thank Emily Ekle (Acquisitions Editor), Liz Palumbo (Associate Managing Editor), Timothy Hiscock (Executive Editor) and Brian Scanlan (President) from Thieme Medical Publishers for all their assistance and encouragement during the journey in making this textbook possible.

Finally, we would like to the thank the reader for taking time from his/her busy schedule to gain insight and improve his/her clinical skills as a result of reading and absorbing all the incredible clinical knowledge that the 68 authors placed between the covers of this textbook.

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Part 1

Hearing Disorders

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1 Posttraumatic Stress Disorder Mimics Auditory Processing Disorder

Michelle Arnold, Theresa Chisolm, Paula J. Myers, and Gabrielle H. Saunders

This case report discusses management of a patient with significant functional hearing concerns following multiple blast exposures, closed blunt head injury, and confirmed mild traumatic brain injury (mTBI) during deployment while serving in the United States Army.

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1.1 Clinical History and Description

MM is a 26-year-old male United States Army veteran who completed two tours of duty during Operation Iraqi Freedom (OIF) between 2007 and 2008. During his deployments, MM was stationed in a theater of combat and sustained no fewer than 25 blast exposures. The most severe event he reported was caused by a rocket-propelled grenade (RPG), which hit the sandstone building above where MM was standing and destroyed most of the structure. MM had been about to enter the building when the RPG detonated. A large piece of sandstone hit MM on the head and knocked him off his feet. As a result of the blast, MM experienced loss of consciousness (LOC) for approximately 1 minute. On regaining consciousness, MM immediately noticed constant bilateral tinnitus. Additionally, MM reported feeling alteration of consciousness for 4 to 5 minutes following the impact and experienced blurred vision and peripheral vision loss that lasted for approximately 20 minutes.

During the same 2007 deployment, MM was exposed to a blast from an improvised rocket-assisted mortar (IRAM), which was fired directly behind him as he was entering a secure federal police compound in Baghdad in an armored vehicle. After the IRAM hit, MM was thrown from the vehicle by the blast and landed facedown. MM lost consciousness for less than 2 minutes and felt alteration of consciousness that lasted for a few minutes. MM also reported that his hearing was “dulled” for the next 12 hours.

In addition to these major incidents, MM sustained 18 to 24 improvised explosive device (IED) exposures while on vehicle convoys during his 2007 and 2008 OIF deployments. MM was wearing his protective Kevlar helmet for all vehicle exposures, which provides substantial protection from blast overpressure. However, MM also reported that he hit the side of his head on the vehicle frame upon impact of the blast wave on multiple occasions. MM did not report use of hearing protection for any blast exposure, and was not wearing any protective head gear for the RPG or IRAM exposures when he was not in a vehicle.

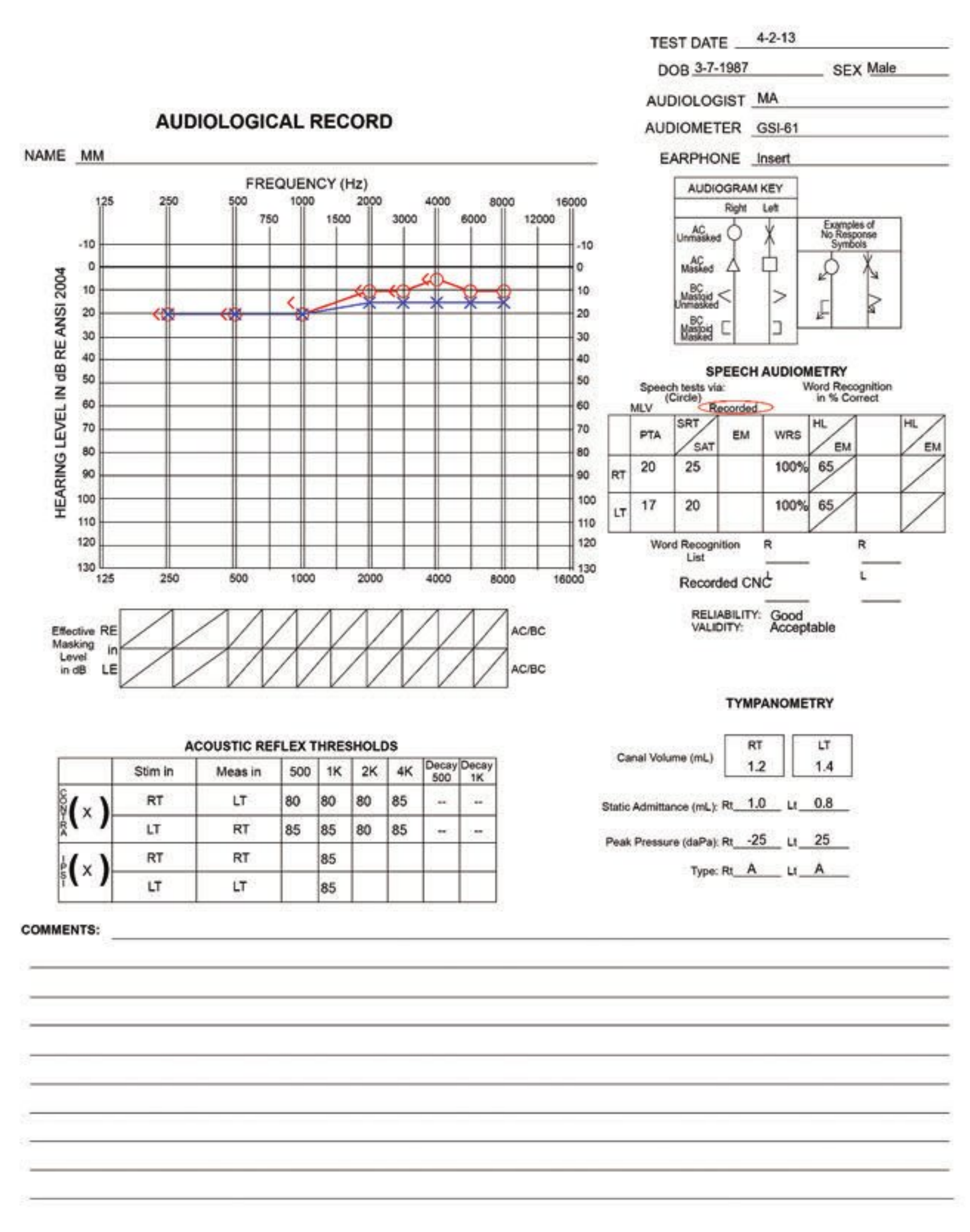
MM planned to use his GI Bill benefits following discharge to attend college. MM experienced no postconcussive symptoms prior to his reported blast exposures that occurred during combat. Following deployment, MM presented to his local Veterans’ Administration (VA) audiology clinic reporting bilateral hearing loss, constant bilateral tinnitus, difficulty hearing in quiet and noise, dizziness/imbalance, memory loss, irritability, sensitivity to noise (i.e., hyperacusis), and problems sleeping. MM arrived at the audiology clinic with a previous diagnosis of posttraumatic stress disorder (PTSD). Further review of his army medical records revealed an abnormal computed tomographic (CT) scan that was performed following the incident in which MM was struck by sandstone. The CT scan displayed intracranial hematomas, which assisted his physician in confirming the presence of mTBI.

1.2 Audiological Testing

Conventional audiological examination included immittance audiometry, distortion-product otoacoustic emissions (DPOAEs), pure-tone and speech audiometry. Pure-tone audiometry revealed bilateral slight hearing loss at 250 to 1000 Hz and rising to be within normal limits at 2000 to 8000 Hz. Speech recognition thresholds (SRTs) revealed a mild loss in the ability to receive speech in the right ear and a slight loss in the ability to receive speech in the left ear. Word recognition scores (WRSs) indicated normal ability to receive speech bilaterally (Fig. 1.1). Tympanometry revealed ear canal volume (mL), peak admittance (mL), and middle ear pressure (daPa) that were within the normal range bilaterally. Contralateral acoustic reflex thresholds (ARTs) were within normal limits at 500 to 4000 Hz. Ipsilateral ARTs at 1000 Hz were within normal limits at 1000 Hz, and acoustic reflex decay was negative at 500 Hz and 1000 Hz bilaterally. MM’s screening revealed a score of 28 on the Hearing Handicap Inventory for Adults, Screening Version (HHIA-S), consistent with a severe self-perceived hearing handicap. Although the results from his audiological examination revealed essentially normal hearing bilaterally, MM continued to exhibit problems with auditory memory as well as difficulties hearing and understanding in diverse listening environments. MM’s subjective report of listening difficulties along with his HHIA-S score indicated a referral for auditory processing testing and treatment to address his functional communication concerns.

1.3 Questions to the Reader

1. What factors in MM’s history may be contributing to the listening difficulties he reports despite normal bilateral peripheral hearing?
2. What additional testing could be recommended for MM?
3. What types of interventions may be appropriate for this veteran?



FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols

AUDIOLOGICAL RECORD

NAME MM

TEST DATE 4-2-13

DOB 3-7-1987 SEX Male

AUDIOLOGIST MA

AUDIOMETER GSI-61

EARPHONE Insert

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	20	25		100%	65		
LT	17	20		100%	65		

Word Recognition List R R

Recorded CNC L L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.4

Static Admittance (mL): Rt 1.0 Lt 0.8

Peak Pressure (daPa): Rt -25 Lt 25

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	80	80	80	85	--	--
	LT	RT	85	85	80	85	--	--
IPSI (X)	RT	RT		85				
	LT	LT		85				

COMMENTS:

Fig. 1.1 Audiological results.

1.4 Discussion of Questions to the Reader

1. What factors in MM's history may be contributing to the listening difficulties he reports despite normal bilateral peripheral hearing?

   It is possible that the listening difficulties MM reported (e.g., trouble hearing in quiet and noise, sensitivity to noise, problems with auditory memory) resulted from poor concentration and memory associated with PTSD. Many veterans exposed to heavy combat are later diagnosed with PTSD following discharge. PTSD symptoms can mimic postconcussive symptoms, even in individuals who have no history of head injury. MM also presented with a confirmed mTBI, likely due to the blunt traumas he experienced during his tours of duty. mTBI can result in auditory deficits, particularly difficulties hearing and understanding in noise, poor auditory memory, and deficits in executive functioning, such as decision making and concentration.
2. What additional testing would you recommend for MM?

   In addition to assessment of MM's functional hearing abilities and auditory processing skills, MM was referred for a comprehensive vestibular evaluation that included ocular motor testing, positional testing, bithermal caloric testing, and rotational chair evaluation to assess his reported dizziness. All vestibular tests were within normal limits.
3. What types of interventions may be appropriate for this veteran?

   Based on MM's history and reported symptoms, MM may benefit from an FM system with a low-gain receiver such as the Phonak iSense (Phonak U.S., Warrenville, IL) coupled with a handheld transmitter, such as the Phonak ZoomLink+or SmartLink+(□ Fig. 1.2). This listening device would be appropriate for use in a classroom environment and may be especially helpful to MM given his desire to begin taking college courses. MM may also benefit from counseling regarding management of his hyperacusis and tinnitus. Progressive tinnitus management (PTM) would be a particularly appropriate management approach because it includes group and individualized counseling that focuses on reducing negative reactions to tinnitus and loud sounds. MM's VA audiologist referred him to group PTM sessions to address his report of tinnitus. MM noted that the amount of attention and time

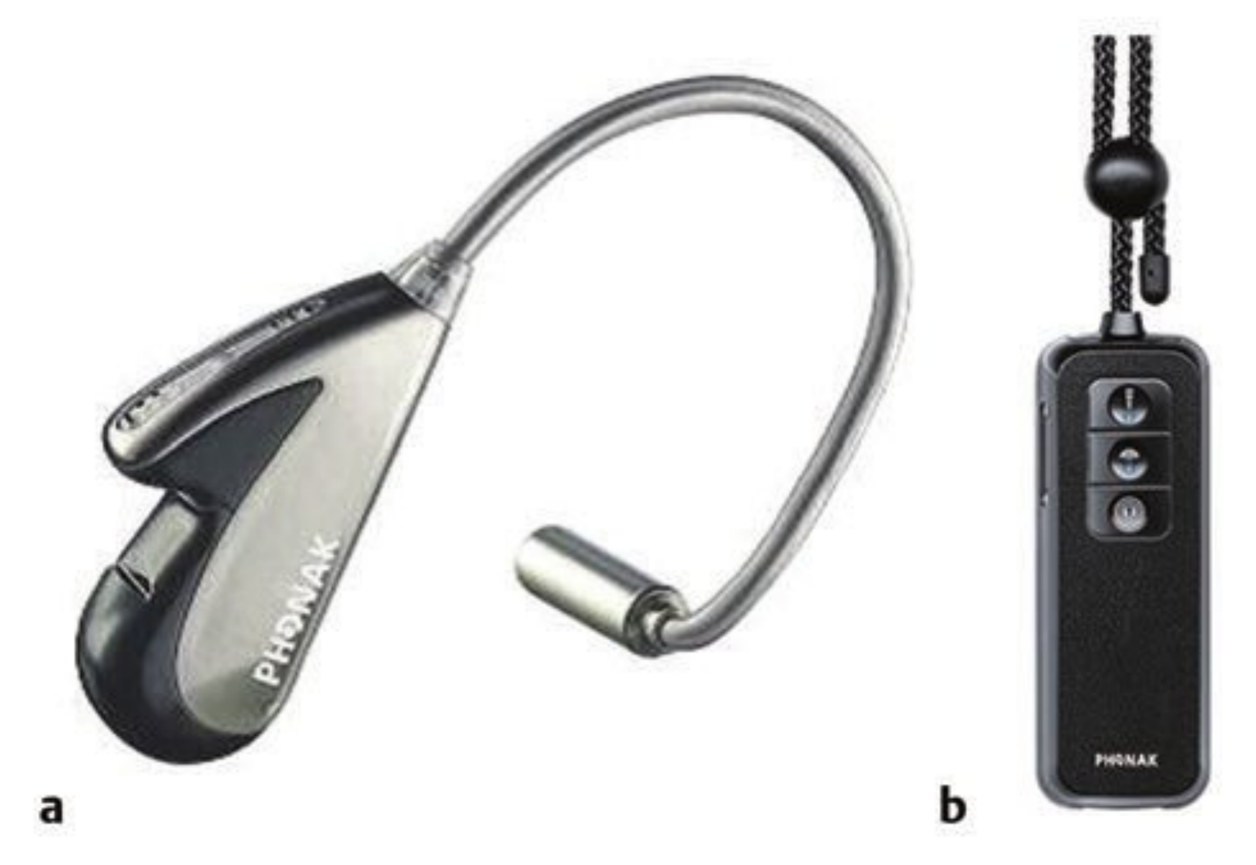


Fig. 1.2 (a) Phonak iSense Micro and (b) Phonak ZoomLink+.

he spent worrying about his tinnitus was greatly reduced following his participation in two PTM sessions. For more information about PTM, see http://www.ncrar.research.va.gov/Education/Documents/TinnitusDocuments/Index.asp.

1.5 Additional Testing

Tests examining MM's functional hearing ability and communication concerns, as well as tests of auditory processing, were administered at a follow-up visit with his VA audiologist. Testing included the Words-in-Noise (WIN) test, the Adaptive Test of Temporal Resolution (ATTR), and the Listening in Spatialized Noise–Sentences (LiSN-S) test. See Table 1.1 for a summary of auditory processing tests, normative values, and MM's results.

The WIN is a speech recognition in noise task that determines the signal to noise ratio (SNR) at which an individual can recognize and correctly repeat 50% of the words. The WIN test uses monosyllables presented in multitalker babble at varying SNRs, starting with a more favorable SNR of +24 dB and ending with a less favorable SNR of 0 dB. MM's score on the WIN test before audiological treatment was 18.8 dB, which is considered a severe SNR hearing loss.

Table 1.1 Objective auditory processing test results

Test	Description	MM's results
WIN	Speech recognition in noise task that determines the signal to noise ratio (SNR) at which an individual can recognize and correctly repeat 50% of the words	18.8 dB SNR (severe SNR loss) (Norm: ≤ 6.0 dB SNR)
ATTR	Gap detection task that assesses temporal resolution using an adaptive paradigm	Within-channel gap detection threshold = 54.5 msec (Norm: 3.0 msec; SD: 0.8 msec)
LiSN-S	Auditory stream segregation task that assesses speech recognition thresholds (SRTs) in noise, as well as talker and spatial listening advantage using target sentences and distracting speech	Low-cue SRT: 14.6 dB (Norm: − 1.1 dB; SD: 0.9 dB) High-cue SRT: 14.2 dB (Norm: − 13.2 dB; SD: 3.0 dB) Talker Advantage: − 0.4 dB (Norm: 7.1 dB; SD: 2.5 dB) Spatial Advantage: − 0.2 dB (Norm: 10.6 dB; SD: 2.2 dB) Total Advantage: 0.5 dB (Norm: 12.2 dB; 2.5 dB)

Abbreviations: ATTR, Adaptive Test of Temporal Resolution; LiSN-S, Listening in Spatialized Noise–Sentences test; SD, standard deviation; WIN, Words-in-Noise test.

The ATTR is an assessment of temporal resolution that measures binaural gap detection; (i.e., the smallest gap a patient is able to detect in noise). Gap detection tasks involve listening to two or more bursts of noise, one of which includes a silent interval embedded in the noise. The ATTR uses an adaptive format in which gaps as small as 1 msec are embedded in diotic noise centered around 1000 Hz. Using a two-alternative forced-choice paradigm, the patient is required to select the interval that contained the gap. If the patient selects incorrectly, the following interval contains a longer gap, and if the patient selects correctly, the following interval contains a shorter gap. Performance on measures of gap detection is thought to be related to processing of degraded speech, such as speech in noise. MM's binaural gap detection threshold was 54.5 msec.

The LiSN-S test is an auditory stream segregation task that assesses speech recognition thresholds (SRTs) in noise, as well as talker and spatial listening advantage using target sentences and distracting speech. The sentences and distracting speech are presented under headphones and are perceived by the listener to be located at different areas in space through the use of head-related transfer functions. Four conditions are tested: (1) target sentences and distracting speech are spoken by different talkers with target sentences arriving from 0° azimuth and distracting speech arriving from ± 90° azimuth (considered a "high-cue SRT"); (2) target sentences and distracting speech are spoken by the same talker with target sentences arriving from 0° azimuth and distracting speech arriving from ± 90° azimuth; (3) target sentences and distracting speech are spoken by different talkers with target sentences arriving from 0° azimuth and distracting speech arriving from 0° azimuth; and (4) target sentences and distracting speech are spoken by the same talker with target sentences arriving from 0° azimuth and distracting speech arriving from 0° azimuth (considered a "low-cue SRT"). By comparing differences in performance on conditions, a talker advantage and a spatial advantage can be computed for a particular listener. A talker advantage is seen as an improvement in performance when the talker of the target sentences is different than the talker of the distracting speech (i.e., Conditions 1 and 3 compared to Conditions 2 and 4). A spatial advantage is seen as an improvement in performance when the distracting speech is arriving from ± 90° azimuth compared with 0° azimuth. MM's low-cue SRT was 14.6 dB and his high-cue SRT was 14.2 dB. MM's talker advantage was − 0.4 dB and his spatial advantage was − 0.2 dB.

1.6 Diagnosis and Recommended Treatment

Based on MM's test results, he was diagnosed with auditory processing disorder (APD). Per the guidelines of the American Speech, Language and Hearing Association, a positive diagnosis of APD is applicable when an individual scores outside two standard deviations below the mean on two or more tests of auditory processing. MM scored outside two standard deviations on each of the three tests of auditory processing ability that were administered (see Table 1.1). MM subsequently received counseling from his audiologist that focused on effective communication strategies, such as facing a speaker when MM was in a conversation, reducing background noise as much as possible when in a conversation, and sitting in the front row during his college courses. Also, MM received informational counseling regarding hearing and speech recognition and hearing conservation strategies. MM's audiologist provided MM with several pairs of disposable ear plugs to use when in excessive noise. MM and his audiologist completed a Client-Oriented Scale of Improvement (COSI). MM was encouraged to generate COSI goals based on his most significant listening challenges. MM's goals were as follows: (1) listening in social situations with less effort and fatigue; (2) listening and attending to college lectures; and (3) feeling less anxious in noisy listening environments, such as sports bars and restaurants. Between his initial appointment at his local VA and a follow-up appointment approximately 12 weeks later, MM received counseling through his local VA Mental Health Services to address his PTSD symptoms.

1.7 Outcome

Twelve weeks after counseling, improvements in performance were seen on the WIN test, the ATTR, and the LISN-S test (see Table 1.2); however, performance on these measures was still below normal (outside two standard deviations from mean performance for adults with normal peripheral hearing sensitivity). MM reported some subjective improvements anecdotally to his audiologist, but none were reflected in his responses on his follow-up COSI. In terms of his listening goals, MM stated that his anxiety and avoidance were significantly reduced, but that the counseling he had received from the audiologist was not responsible for his improvements. He attributed the improvements to the therapy he had received through VA Mental Health Services. It is also possible that the improvements in performance on the auditory processing tests (WIN, ATTR, and LISN-S) could be attributed to improved concentration and attention following MM's mental health counseling.

Table 1.2 Objective auditory processing follow-up test results

Test	Description	MM's results
WIN	Speech recognition in noise task that determines the signal to noise ratio (SNR) at which an individual can recognize and correctly repeat 50% of the words	15.6 dB SNR (severe SNR loss) (Norm: ≤ 6.0 dB SNR)
ATTR	Gap detection task that assesses temporal resolution using an adaptive paradigm	Within-channel gap detection threshold = 15.5 msec (Norm: 3.0 msec; SD: 0.8 msec)
LiSN-S	Auditory stream segregation task that assesses speech recognition thresholds (SRTs) in noise, as well as talker and spatial listening advantage using target sentences and distracting speech	Low-cue SRT: 10.6 dB (Norm: − 1.1 dB; SD: 0.9 dB) High-cue SRT: 8.2 dB (Norm: − 13.2 dB; SD: 3.0 dB) Talker advantage: 1.4 dB (Norm: 7.1 dB; SD: 2.5 dB) Spatial advantage: 5.6 dB (Norm: 10.6 dB; SD: 2.2 dB) Total advantage: 4.5 dB (Norm: 12.2 dB; 2.5 dB)

Abbreviations: ATTR, Adaptive Test of Temporal Resolution; LiSN-S, Listening in Spatialized Noise–Sentences test; SD, standard deviation; WIN, Words-in-Noise test.

MM was then offered an FM trial as well as use of computerized auditory training with the Brain Fitness Program (Posit Science Corp., San Francisco, CA) to further address his auditory deficits. The Brain Fitness Program (BFP) consists of six exercises designed to enhance temporal processing and auditory working memory skills through intensive practice listening to temporally altered signals and changing memory loads. The exercises are adaptive, such that task difficulty automatically adjusts based on the user's performance. The reader should note that as of 2012, the BFP program is now incorporated into Posit Science online BrainHQ program, which is available as a month subscription. For more information on Brain Fitness or BrainHQ, see http://www.positscience.com. MM reported using the FM system for about 3 hours a day and completed 9 of the 20 recommended training sessions. MM reported that he found the BFP and the FM listening system to be helpful and that the combination of mental health and audiological services he received had resulted in significant improvements in his daily quality of life.

1.8 Key Points

1. Many thousands of Operation Iraqi Freedom and Operation Enduring Freedom veterans are presenting in VA clinics nationwide with a wide array of auditory and vestibular deficits following tours of duty. Many of these patients are also presenting to university and hospital audiology clinics and private practices. Conventional audiological examination on many of these patients reveals normal peripheral hearing sensitivity, yet auditory complaints continue to persist. Patients with a history of mTBI who present with persistent functional auditory deficits despite normal audiological results should be referred for further assessment and treatment of their functional hearing and communication difficulties. Part of this assessment may include auditory processing testing.
2. The reader should be aware that it is sometimes necessary to refer patients for mental health services, and these services are very important. For MM, audiological intervention alone may not have improved his listening deficits, but the combined audiological and mental health services resulted in a positive outcome.

Suggested Reading

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2 Use of a Stent in a Collapsing Exterior Ear Canal

Marshall Chasin and Anil Katyal

CP is a 68-year-old woman who has bilateral polychondritis leading to a collapsing of the cartilaginous portion of her ear canals. The presence of collapsing ear canals has ramifications for accurate audiological examinations. This case report highlights a simple solution to solve this problem while improving CP's communication ability.`

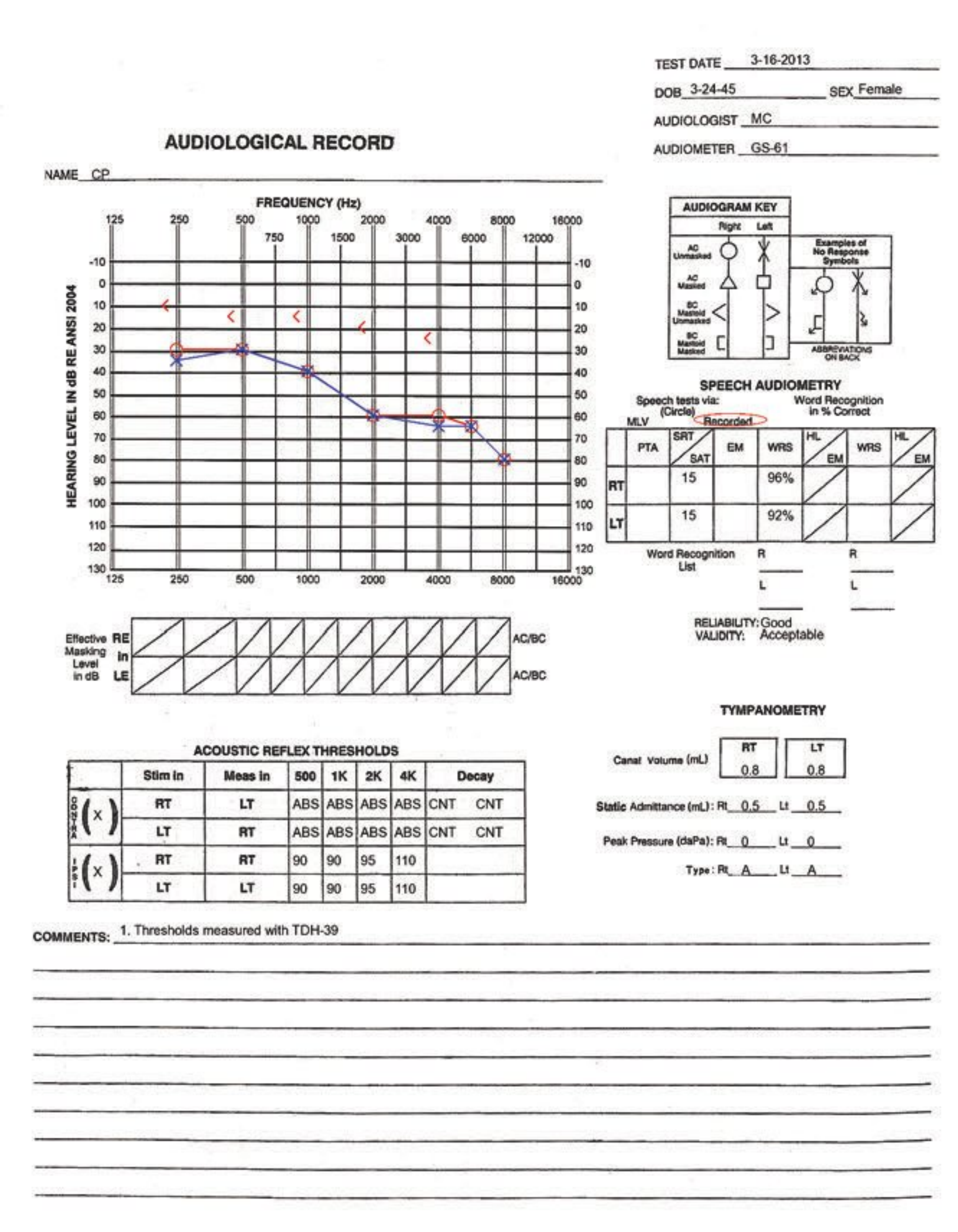
2.1 Clinical History and Description

CP was seen with reports of a progressive hearing loss that manifested itself in noisy and other adverse listening environments. She expressed no difficulty with one-to-one communication. There were no reports of ear pain or ear infections. CP reported she could hear well, but people mumbled. CP reported that she was diagnosed with polychondritis and that hearing had decreased even further in the last several years.

Polychondritis or relapsing polychondritis is an inflammatory disease of cartilage, typically involving the cartilage of the ear, nose, larynx and upper respiratory tract, thorax, and joints. This disease typically affects middle-aged women. Patients may initially present with beefy, tender red swellings of the cartilage of the external ear or other affected cartilaginous structures, along with other symptoms, including hoarseness and tenderness over the larynx, joint pain, malaise, and fever. The ongoing intermittent bouts of inflammation can lead to cartilage destruction, granulation tissue formation, and fibrosis. Although polychondritis usually affects the pinnae of the ears, any extension to the outer cartilaginous portion of the ear canal can eventually lead to canal cartilage softening and destruction, leading to loss of support and collapse of the outer external canal. Such events would lead to a conductive hearing loss in the affected ear.

2.2 Audiological Testing

A comprehensive audiological examination (Fig. 2.1) was performed with a suspicion of presbycusis. Immittance audiometry indicated normal ear canal volume, static compliance and



AUDIOLOGICAL RECORD

TEST DATE 3-16-2013
DOB 3-24-45 SEX Female
AUDIOLOGIST MC
AUDIOMETER GS-61
NAME CP

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT		15		96%			
LT		15		92%			

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay	
CONTRA (X)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (X)	RT	RT	90	90	95	110		
	LT	LT	90	90	95	110		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.8	0.8

Static Admittance (mL): Rt 0.5 Lt 0.5
Peak Pressure (daPa): Rt 0 Lt 0
Type: Rt A Lt A

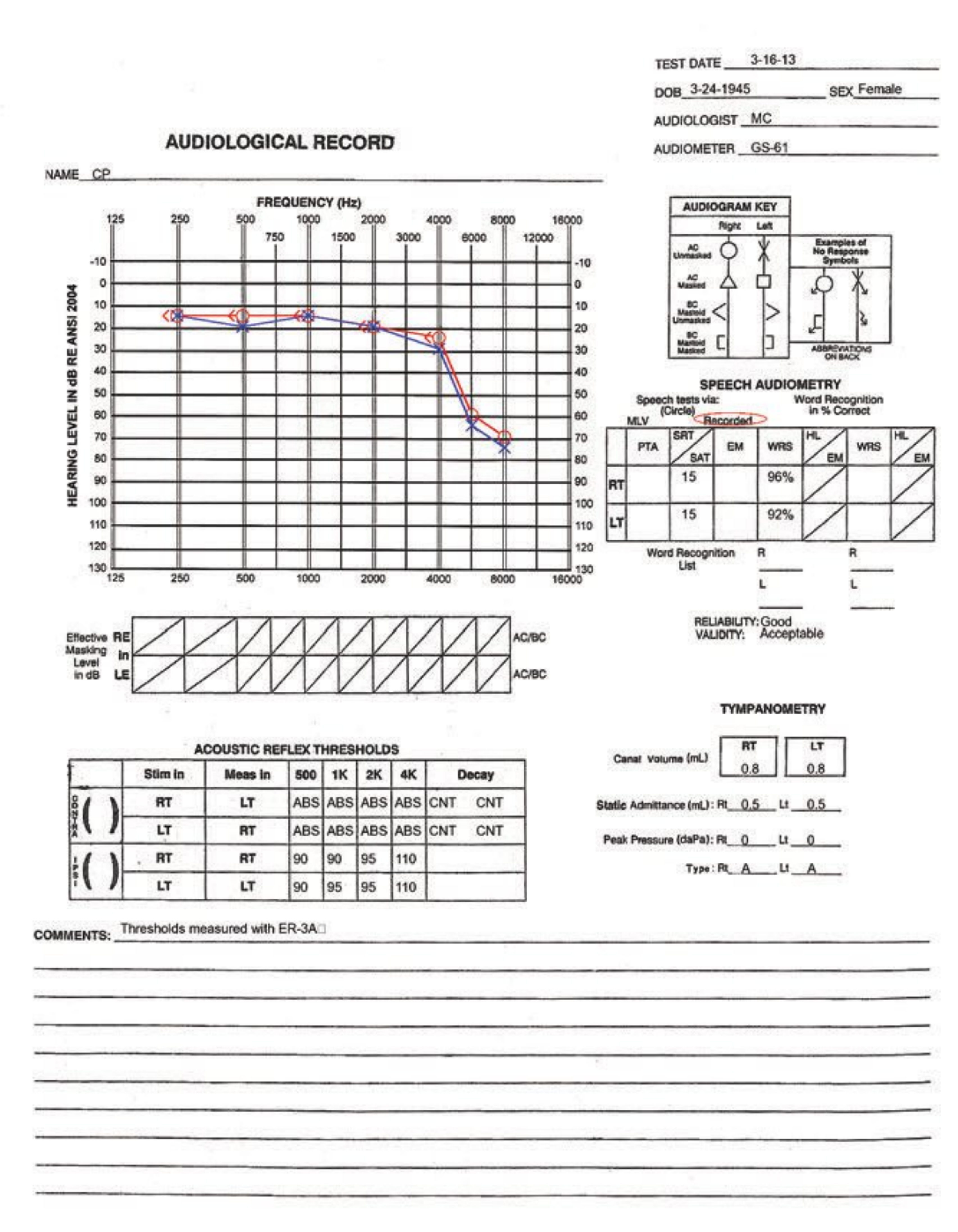
COMMENTS: 1. Thresholds measured with TDH-39

Fig. 2.1 Audiological examination reporting the results using TDH-39 earphones. Note the significant air–bone gaps at 250 to 4000 Hz with normal tympanograms and absent acoustic reflex thresholds to contralateral stimulation at 500 to 4000 Hz.

middle ear pressure bilaterally, but absent contralateral acoustic reflex thresholds (ARTs) bilaterally at 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz. Ipsilateral ARTs were present bilaterally although slightly elevated at 4000 Hz. Using TDH-39 earphones, audiological examination revealed a mild to moderate–severe high-frequency conductive hearing loss at 250 to 1000 Hz and a slight mixed hearing loss at 2000 to 4000 Hz. These results revealed a 35 dB air–bone gap in the higher-frequency region and a 15 to 20 dB air–bone gap at 250 to 500 Hz. Speech recognition thresholds (SRTs) were consistent with the pure-tone average and indicated normal ability to receive speech bilaterally. Word recognition scores (WRSs) revealed normal ability to receive speech bilaterally (96% in the right ear and 92% in the left ear). Although not shown, otoacoustic emission (OAE) testing revealed responses that were within the normal range to 2000 Hz and were present but slightly reduced at 4000 Hz.

Because of the contradictory nature of the initial audiological examination (i.e., conductive-mixed hearing loss with normal tympanograms and ipsilateral ARTs and OAEs), air conduction thresholds were repeated using ER-3A insert earphones (Etymotic Research, Inc., Elk Grove Village, IL) with small-sized foam connectors (Fig. 2.2). CP was asked to keep her jaw open during the insertion of the foam ear tips since this would allow the condyle of her mandible to slide forward, which would temporarily increase the anterior-posterior dimension of her ear canals. This procedure would allow the foam inserts to be placed more deeply in her ear canals for more reliable testing. As a result of the repeated examination, air conduction thresholds were now found to be within 5 dB of the original bone conduction thresholds, thereby resolving the air–bone gap. It was explained to CP that her hearing was normal for her age and that any contradictory test results were probably due to the audiometric earphones collapsing her ear canals causing a test artifact.

Several days later, however, CP again contacted the office asking for an appointment to retest her hearing because she was still experiencing significant communication difficulties in noisy locations. Air conduction thresholds were once again assessed with insert earphones, and again her hearing was found to be normal except for a mild high-frequency sensorineural hearing loss at 4000 Hz and above.



AUDIOLOGICAL RECORD

TEST DATE 3-16-13
DOB 3-24-1945 SEX Female
AUDIOLOGIST MC
AUDIOMETER GS-61
NAME CP

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT		15		96%			
LT		15		92%			

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay	
CONTRA ( )	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI ( )	RT	RT	90	90	95	110		
	LT	LT	90	95	95	110		

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	0.8	0.8

Static Admittance (mL): Rt 0.5 Lt 0.5
Peak Pressure (daPa): Rt 0 Lt 0
Type: Rt A Lt A

COMMENTS: Thresholds measured with ER-3A

Fig. 2.2 Audiological examination reporting the results using ER-3A insert earphones (Etymotic Research, Inc.). Note that the air–bone gaps are no longer present.

2.3 Questions to the Reader

1. How can ipsilateral ARTs be present if there is a significant air–bone gap?
2. How can OAEs be measured with the presence of significant air–bone gaps and the magnitude of the hearing loss found during the initial audiological examination of CP?
3. Which phonemes would CP have difficulty recognizing and what contribution would these phonemes provide to the Speech Intelligibility Index (SII) for an English talker?
4. Why does a collapsed canal result in a "high-frequency" conductive hearing loss?

2.4 Discussion of Questions to the Reader

1. How can ipsilateral ARTs be present if there is a significant air–bone gap?
   The testing of ipsilateral ARTs uses a probe and tip that forces the outer ear canal open during testing. Therefore, any collapsing of the ear canal would not have been observed. This was not the case when using the TDH-39 circumaural headphone to assess contralateral ARTs as well as the initial audiological air conduction thresholds. Differences in this case were due to the mechanical configuration of one stimulus delivery device (insert probe) as compared with another (TDH-39 earphones).
2. How can OAEs be measured with the presence of significant air–bone gaps and the magnitude of the hearing loss found during the initial audiological examination of CP?
   OAE testing, like the testing for ipsilateral ARTs, uses a probe and tip that prop the ear canal open during testing. Therefore, the outer ear collapsing canal was remediated for OAE testing as it was for ipsilateral ARTs. The use of the TDH-39 earphones during the audiological evaluation revealed air–bone gaps, but these air–bone gaps were related to the method of stimulus delivery (i.e., TDH-39 earphones) rather than CP's anatomical condition.
3. Which phonemes would CP have difficulty recognizing and what contribution would these phonemes provide to the Speech Intelligibility Index (SII) for an English talker?
   With the hearing loss reported in Fig. 2.1, showing air–bone gaps and high-frequency hearing loss, the SII would be 40%, implying that only 40%of English speech cues were audible. With the ear canal propped open, the audiometric thresholds improved, and there was an associated improvement in the SII to 100%Much of the contribution to the SII comes from the higher frequency siblants such as /s, z, f,.../..
4. Why does a collapsed canal result in a "high-frequency" conductive hearing loss?
   The ear canal functions as a quarter wavelength resonator (closed at the medial tympanic membrane end and open at the lateral meatal end). This is similar to the "1000 Hz" resonance in the frequency response of behind-the-ear (BTE) hearing aids as well as during the articulation of the vowel [a] as in father. The natural quarter wavelength resonance of an adult ear canal is approximately 2700 Hz, with lower frequencies having a smaller amplitude of the associated standing wave. Because it is the lateral (cartilaginous) portion of the ear canal that collapses, the standing wave has a larger volume velocity (marked with the first vertical line in Fig. 2.3 on the left side) in this region, and the energy associated with this natural high frequency resinance is reduced. In contrast, if the obstruction was adjacent (marked with the second vertical line in Fig. 2.3) but not in contact with the tympanic membrane (on the medial side) there would be minimal effect on the standing wave pattern because the volume velocity is very small in this lateral region.

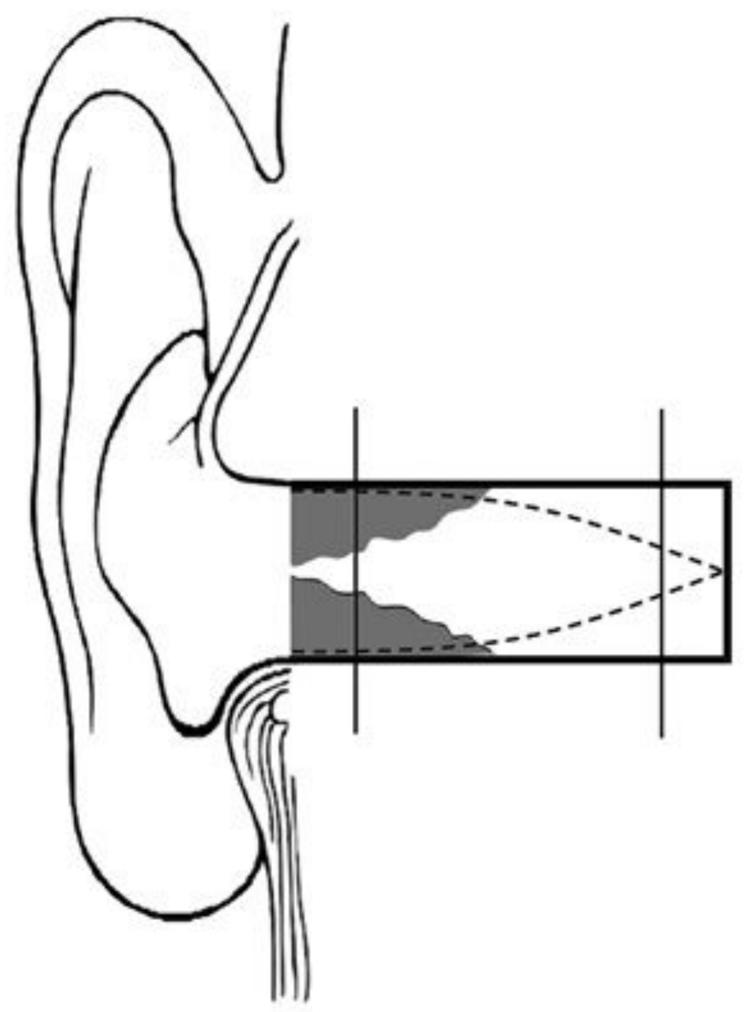


Fig. 2.3 Schematic drawing of the outer ear canal showing a standing wave associated with the quarter wavelength resonance (open at the lateral side and closed at the medial tympanic membrane side). The shaded region reports the presence of the collapsed cartilage found commonly with polychondritis. An obstruction at the second vertical line (on the right side) would have a minimal effect of the destruction of the ear canal resonance, but an obstruction at the first vertical line (on the left side) would have a significant effect. A lateral obstruction at the first vertical line by a collapsing ear canal would result in a high-frequency air–bone gap during audiological assessment.

   In addition to the destruction of the natural resonance the narrowing of the ear canal contributes to the overall impedance. This results in a further lessening of the high frequency energy. The longer wavelength of the lower-frequency signals "perceives" the collapsed canal obstruction as being more acoustically transparent. Acoustically, this is equivalent to stating that acoustic impedance increases with frequency.

2.5 Additional Testing

A real ear probe tube microphone measurement was completed twice to assess CP's real ear unaided response (REUR). One REUR was measured with the ear canal in its collapsed state, and a second measure was made with CP's ear canal propped open with the Lucite stent in place. Fig. 2.4 reports the REUR for the two measures using an input level of 70 dB SPL (Sound Pressure Level) to ensure that the measured results were above the internal noise floor of the real ear measurement equipment. The lower curve reports the REUR with a collapsing ear canal,

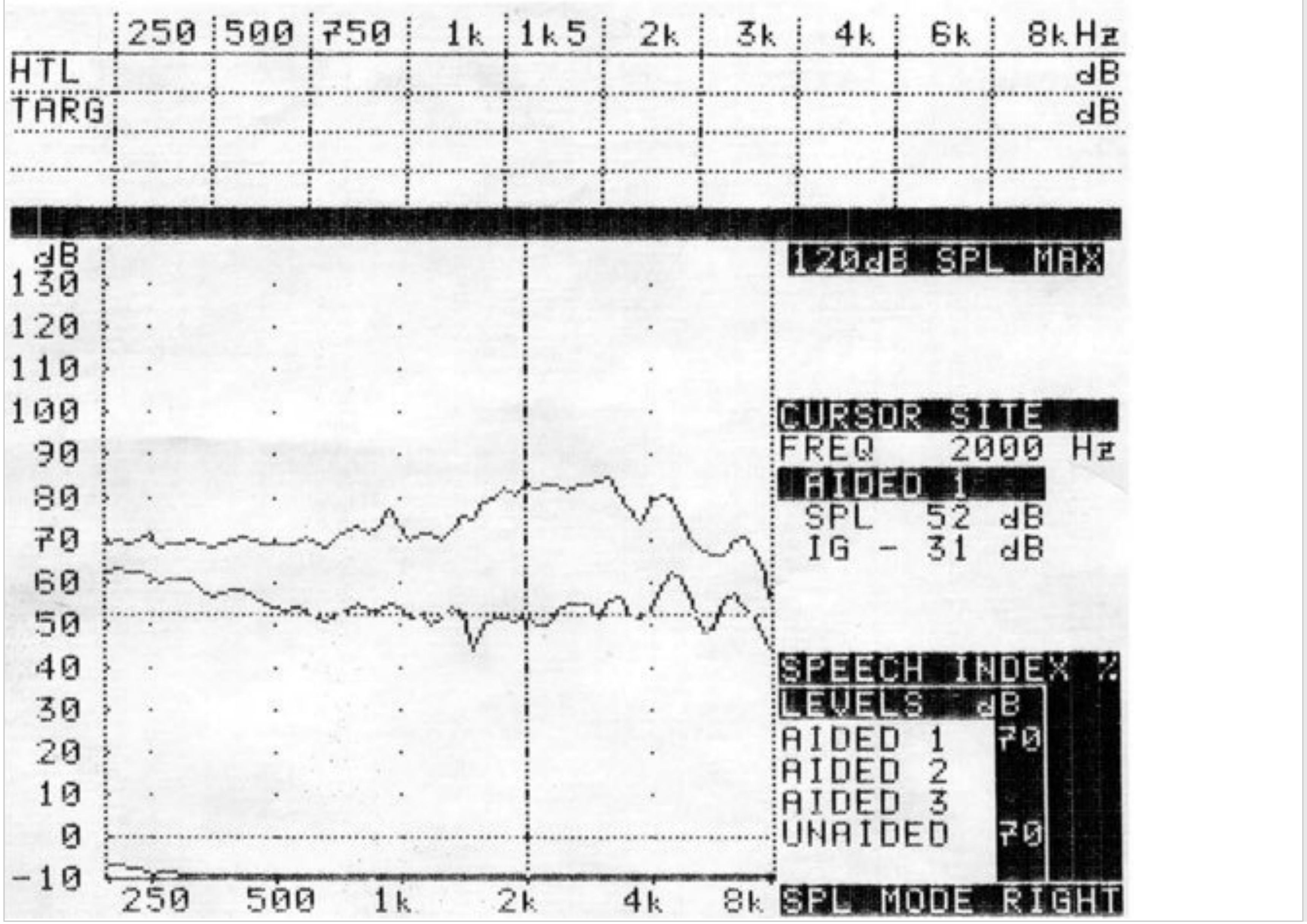


Fig. 2.4 The real ear unaided response (REUR) of a normal ear canal (upper REUR) and one with an obstruction at the meatal (lateral) opening (lower REUR). The lower curve reports the REUR with a collapsing ear canal, and the upper curve reports the REUR with the Lucite stent in position. The upper curve is similar to the REUR of a normal unoccluded ear canal.

and the upper curve reports the REUR with the Lucite stent in position. The upper curve is similar to the REUR of a normal unoccluded ear canal.

2.6 Diagnosis and Recommended Treatment

The use of an ER-3A insert earphone resolved the air–bone gap that was suggestive of a collapsed ear canal; however, due to her polychondritis, this collapsing of both ear canals was present even without the added pressure of the TDH-39 earphones. CP continued to experience a 35 dB conductive overlay at 4000 Hz, which, along with her 25 dB HL bone conduction threshold at 4000 Hz, yielded a 60 dB HL loss of sensitivity for the higher-frequency sibilants of speech.

A custom set of Lucite stents (hollow ear pieces where the external side is in contact with the outer ear canal walls and the interior portion is hollowed out as much as possible) were manufactured bilaterally. These can be ordered from any earmold laboratory by requesting a canal tip style mold that is hollowed out as much as possible given the limitations of the possibly narrow ear canal impression. These stents have a 3 mm inner diameter air hole, which served to prop open her ear canals such that her effective hearing thresholds were consistent with her bone conduction thresholds. The Lucite stents were fitted with small nylon removal cords similar to that of a completely in-the-canal (CIC) hearing aid. During the earmold impression procedure, CP was asked to keep her jaw wide open. This procedure allowed the condyle of her jaw to move forward and away from the anterior wall of the external ear canal. The “collapsing” of the ear canal was therefore minimized while the earmold impressions were made. Also, given the nature of the collapsing ear canal and the diagnosis of polychondritis, CP was referred to an otolaryngologist.

2.7 Outcome

CP was able to carry on her normal activities and now could hear most people, even in many adverse listening environments. CP’s measured SII improved from 40 to 100% The measurement of the SII was accomplished by entering her audiometric information into a real ear measurement system that has the capability of calculating the SII, once with the initial (collapsed canal) audiometric thresholds, and again with the audiometric thresholds when an insert earphone or the Lucite stent was used.

2.8 Key Points

1. Ensure that the mechanical aspects of the method of sound transduction are taken into account during audiological examinations.
2. Collapsed canals typically result in a high-frequency conductive or mixed hearing loss because of the nature of the wavelength sound transmission in the outer ear canal.
3. Present ipsilateral ARTs and normal OAEs are inconsistent with conductive hearing loss.
4. Polychondritis can be a cause of permanent loss of cartilage in the outer ear that can result in partial or complete closure of the ear canal with an associated high-frequency conductive hearing loss.
5. A collapsed canal due to insufficient cartilage support can be propped open by the use of a custom-made mechanical (Lucite) hollow ear piece.

Suggested Reading

McDonald TJ. Manifestations of Systemic Disease in the External Ear. In Cummings CW. ed. Otolaryngology - Head and Neck Surgery: St. Louis, MO: Mosby Publishing, St; 1998: 2996–2997

3 Auditory Neuroma

Susan E. Fulton

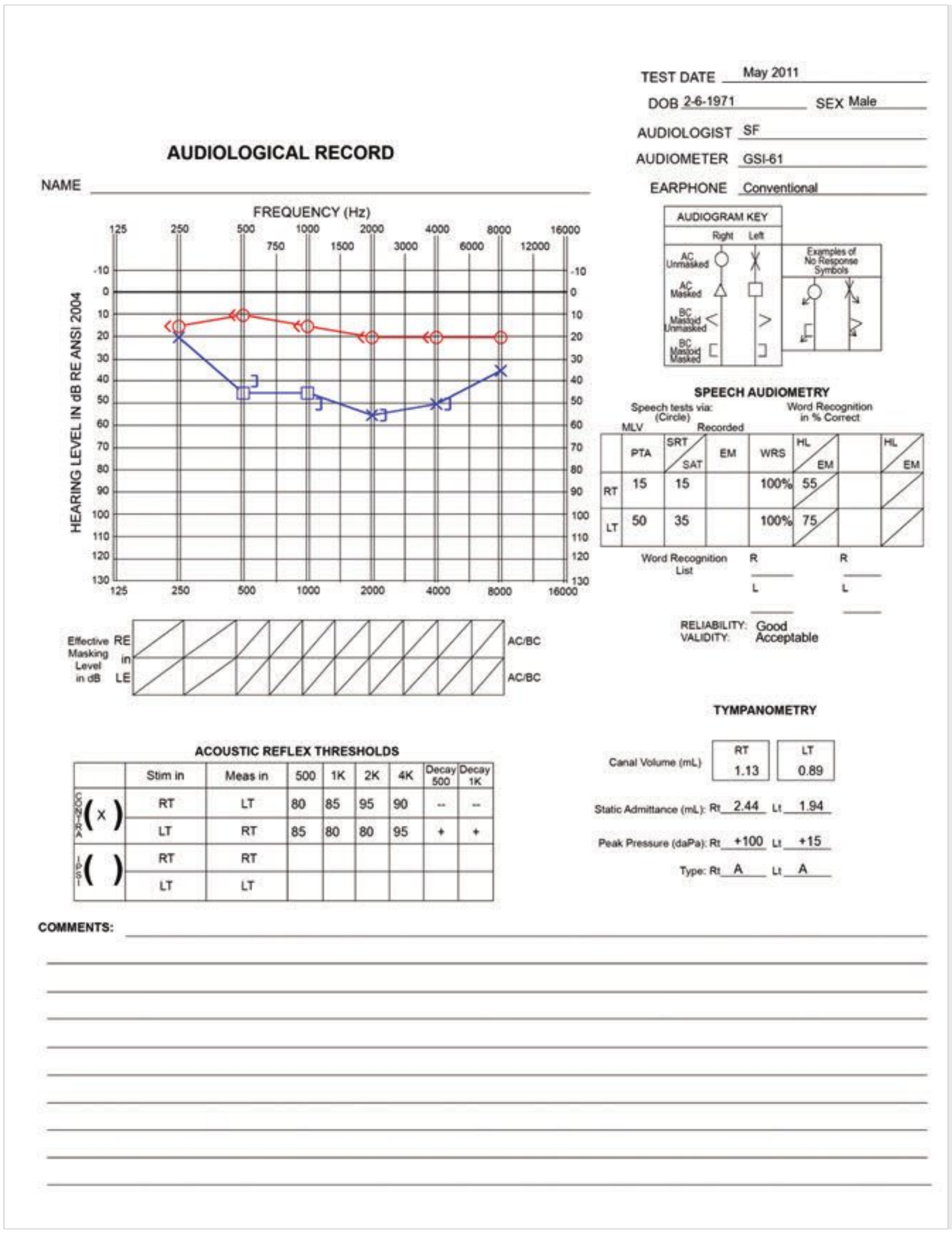
This is a case report of a 42-year-old male who presented with a mild to moderate sensorineural hearing loss, intermittent tinnitus, and aural fullness in the left ear.

3.1 Clinical History and Description

A 42-year-old male was seen for an initial audiological examination. He presented with a report of a gradual hearing loss, intermittent tinnitus, and aural fullness in the left ear. His hearing sensitivity has reportedly deteriorated slowly over the last 2 years. The patient has a history of unprotected noise exposure working as a club promoter for many years and recently has been exposed to machinery noise while working in construction. The patient denies otalgia, otorrhea, or dizziness.

3.2 Audiological Testing

An audiological examination revealed clear external canals bilaterally. Pure-tone thresholds (Fig. 3.1) indicated hearing within normal limits at 250 to 1000 Hz, a slight hearing loss at 2000 to 8000 Hz in the right ear, and a slight to moderate sensorineural hearing loss in the left ear at 250 to 4000 Hz with recovery to a mild sensorineural hearing loss at 8000 Hz. Speech recognition thresholds (SRTs) were at 15 dB HL in the right ear and 35 dB HL in the left ear indicating normal ability to receive speech in the right ear and a mild loss in the ability to receive speech in the left ear. The SRTs were considered to be in good agreement with pure-tone averages (PTAs) for the right ear but were slightly elevated for the left ear. Word recognition scores (WRSs) revealed normal ability to recognize words bilaterally at 40 dB sensation level (SL). Immittance audiometry



TEST DATE May 2011
DOB 2-6-1971 SEX Male
AUDIOLOGIST SF
AUDIOMETER GSI-61
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	15	15		100%	55		
LT	50	35		100%	75		

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.13	0.89

Static Admittance (mL): Rt 2.44 Lt 1.94
Peak Pressure (daPa): Rt +100 Lt +15
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	80	85	95	90	--	--
	LT	RT	85	80	80	95	+	+
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 3.1 Initial audiological examination.

revealed type Ad tympanograms bilaterally, indicative of hypercompliant tympanic membranes with normal middle ear pressure (daPa), normal ear canal volume (mL), and slightly elevated static admittance (mL). Contralateral acoustic reflex thresholds (ARTs) were within normal limits at 500 to 4000 Hz bilaterally. Contralateral reflex decay was negative in the right ear and positive in the left ear at 500 and 1000 Hz. Otoacoustic emissions were absent at 1500 to 6000 Hz bilaterally.

3.3 Questions to the Reader

1. Based on the initial results of the audiological evaluation, what pathology would be most likely?
2. What recommendations should be made for this patient?
3. In that the patient presents with a hearing loss in the left ear, can amplification be recommended at this time?
4. How should the patient be counseled at this time?

3.4 Discussion of Questions to the Reader

1. Based on the initial results of the audiological evaluation, what pathology would be most likely?
   Audiological results indicate a unilateral sensorineural hearing loss. Left-sided aural fullness and tinnitus along with positive left ear contralateral reflex decay suggest possible retrocochlear pathology.
2. What recommendations should be made for this patient?
   Due to suspected left-sided retrocochlear pathology, the patient should be referred to an otologist for further examination. The otologist may order auditory brainstem response (ABR) and/or image testing.
3. In that the patient presents with a hearing loss in the left ear, can amplification be recommended at this time?
   Amplification may be an option in the future. Amplification, however, cannot be fit when a medical condition is suspected, unless a physician provides medical clearance.
4. How should the patient be counseled at this time?
   The audiologist should explain the results of the audiological examination. Suspicion of problems past the cochlea should be discussed. The patient should be counseled to schedule an appointment with an otologist for further medical intervention. The possibility of amplification, pending medical clearance, should be discussed. Ear protection in the presence of loud noise should be recommended.

3.5 Diagnosis and Recommended Treatment

The patient was referred to an otologist. The otologist ordered an ABR test and magnetic resonance imaging (MRI) of the temporal area. The ABR test with a standard stimulus rate revealed normal peak and interpeak latencies in the right ear and delayed peak and interpeak latencies in the left ear. Increased stimulus rate revealed no adverse effects in the right ear and continued delayed peak latencies in the left ear. An MRI scan with and without contrast indicated a 4 × 3 × 2 mm left internal auditory canal lesion suggesting the presence of an acoustic neuroma.

3.6 Outcome

Surgery was performed and a left acoustic neuroma was removed (Fig. 3.2). Fig. 3.2c reveals that the tumor was encapsulated and easily removed. A depression in the auditory nerve was present after the space-occupying lesion (tumor) was removed (Fig. 3.2d). The patient reported dizziness and left-sided facial weakness following surgery. He also experienced a decrease in hearing sensitivity in the left ear. Due to the lingering dizziness, the patient has been unable to continue in

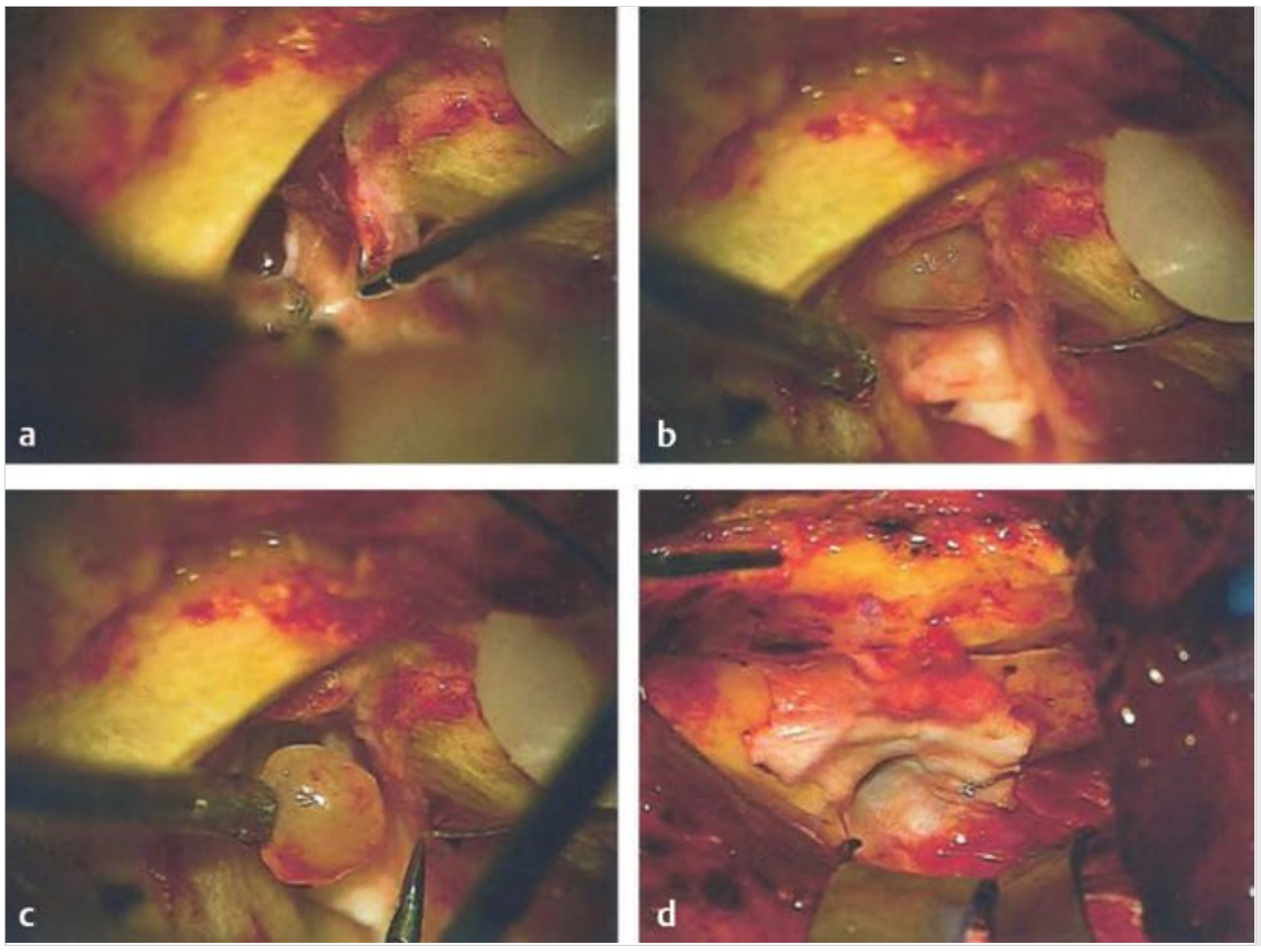
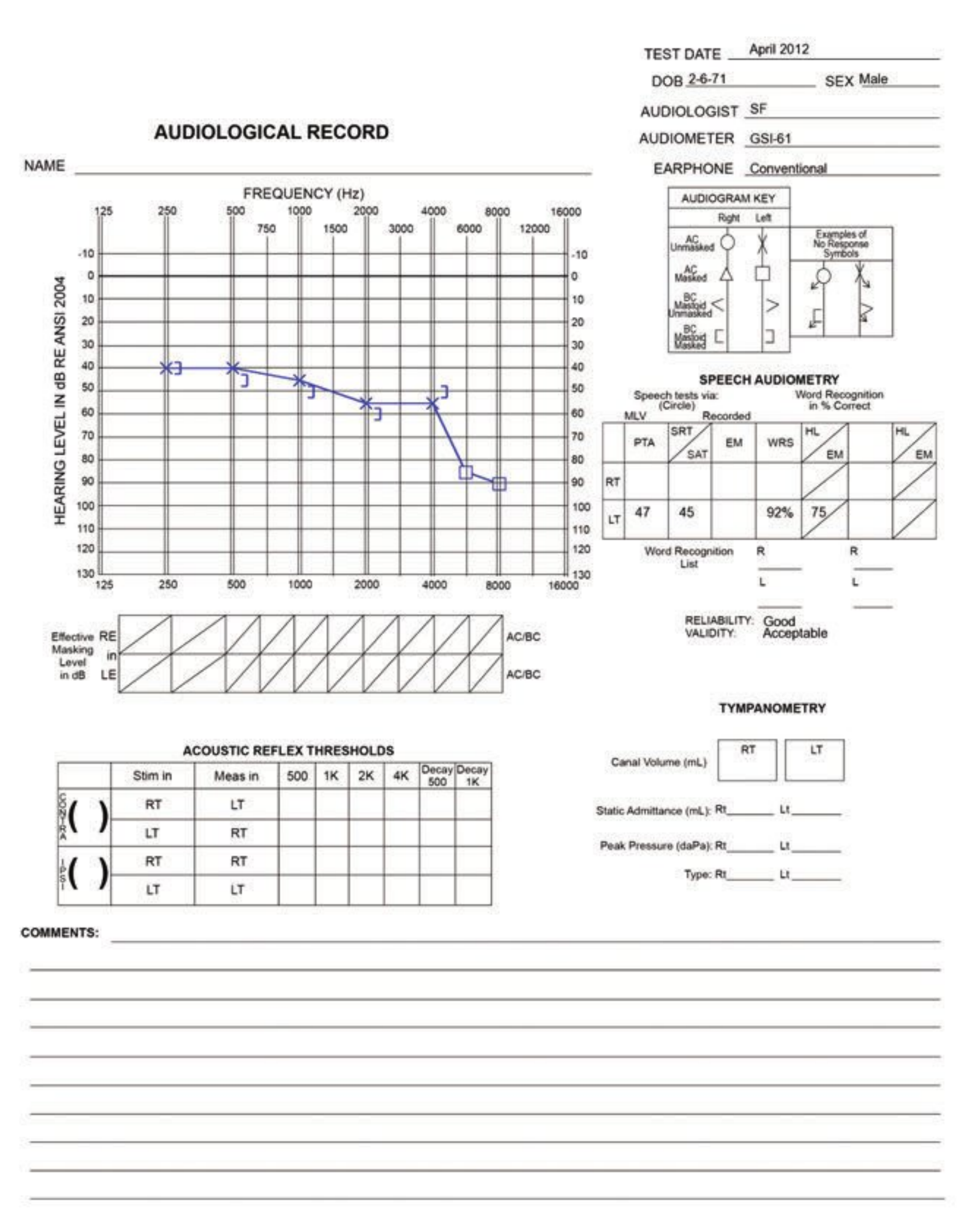


Fig. 3.2 Auditory neuroma excision. (a) Auditory nerve and surrounding tissues. (b) Auditory neuroma location. (c) Removal of auditory neuroma. (d) Site following removal of auditory neuroma.



AUDIOLOGICAL RECORD

NAME

TEST DATE April 2012

DOB 2-6-71 SEX Male

AUDIOLOGIST SF

AUDIOMETER GSI-61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT							
LT	47	45		92%	75		

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt Lt

Peak Pressure (daPa): Rt Lt

Type: Rt Lt

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 3.3 Postoperative audiological examination (left ear testing only) obtained 11 months following initial audiological examination.

his profession as a construction worker. He has received counseling from Vocational Rehabilitation (VR) and is pursuing a new career. A postoperative audiological examination (Fig. 3.3) reveals a significant decrease in hearing sensitivity at 250 to 8000 Hz in the left ear, which has remained stable since surgery.

3.7 Key Points

1. Retrocochlear pathology should be suspected when a patient presents with asymmetrical hearing loss, unilateral tinnitus, and unilateral aural fullness.
2. WRSs may not be affected when an auditory lesion is present.
3. Acoustic reflex decay is an important diagnostic tool in identifying retrocochlear lesions.
4. Auditory and vestibular function may be adversely affected following acoustic neuroma removal.

Suggested Reading

Brooker J, Burney S, Fletcher J, Dally M. A qualitative exploration of quality of life among individuals diagnosed with an acoustic neuroma. Br J Health Psychol 2009; 14: 563–578

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Godey B, Morandi X, Beust L, Brassier G, Bourdinière J. Sensitivity of auditory brainstem response in acoustic neuroma screening. Acta Otolaryngol 1998; 118: 501–504

Schlauch RS, Levine S, Li Y, Haines S. Evaluating hearing threshold differences between ears as a screen for acoustic neuroma. J Speech Hear Res 1995; 38: 1168–1175

Tos M, Stangerup SE, Cayé-Thomasen P, Tos T, Thomsen J. What is the real incidence of vestibular schwannoma? Arch Otolaryngol Head Neck Surg 2004; 130: 216–220

4 Functional Hearing Loss

Susan E. Fulton and Frederick E. Cobb

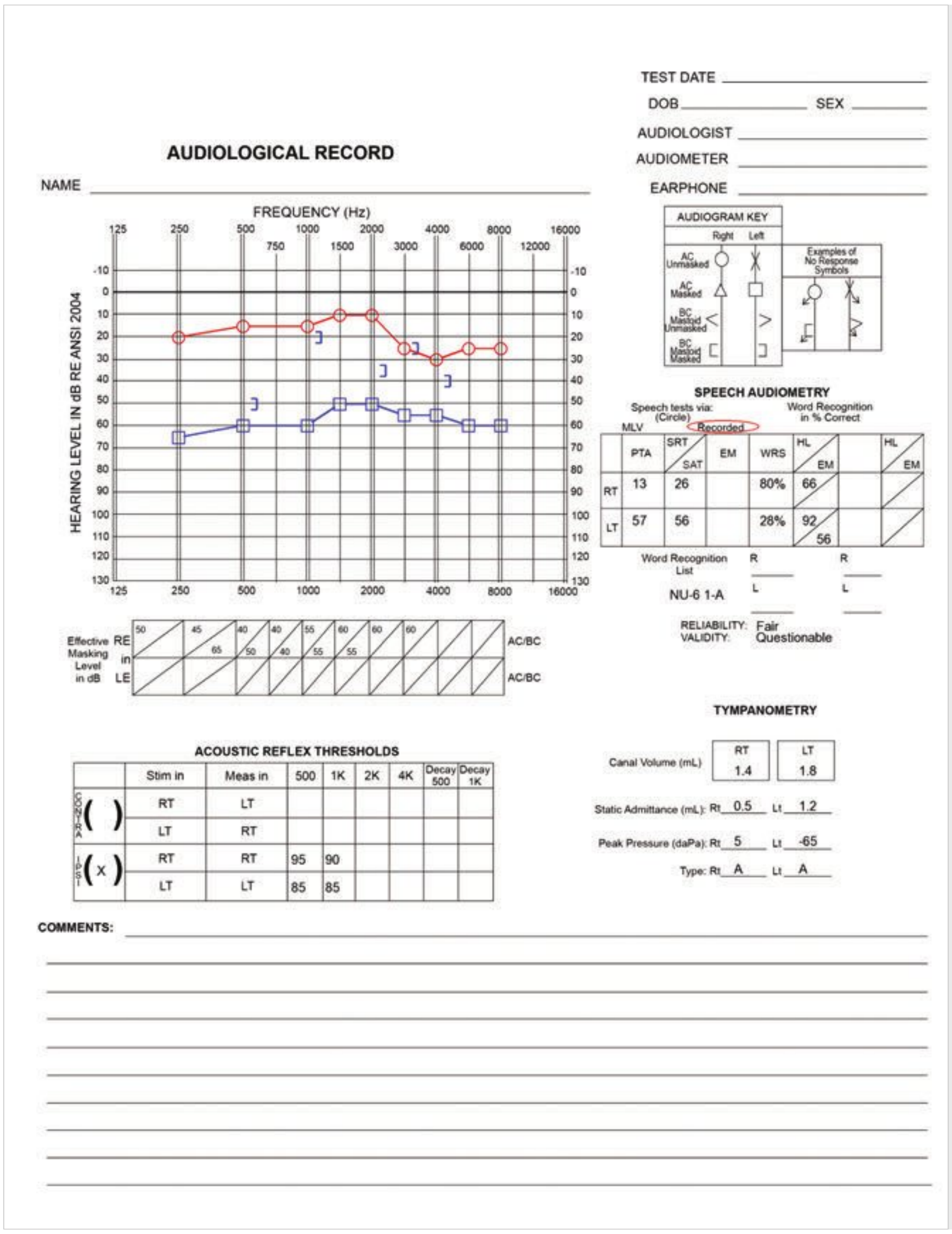
This is a case report of a 63-year-old male who presented with complaints of a hearing loss and constant tinnitus in his left ear. The onset of the hearing loss was reportedly more than 10 years ago.

4.1 Clinical History and Description

A 63-year-old male was seen for an initial audiological examination. He presented with a report of hearing loss and constant tinnitus in the left ear. He had noticed the hearing loss for the past 10 years, and he described the tinnitus as a “wind noise or low hum.” He reportedly consulted a physician at onset, but no medical intervention was recommended. The patient denied otalgia, otorrhea, or dizziness.

4.2 Audiological Testing

An otoscopic observation revealed clear external canals bilaterally. The initial results from the audiological examination (Fig. 4.1) for the right ear revealed pure-tone thresholds indicating a slight hearing loss at 250 and 3000 to 8000 Hz with normal hearing at 500 to 2000 Hz. Results for the left ear revealed a moderately severe mixed hearing loss at 250 to 8000 Hz that is essentially flat in configuration. Speech recognition thresholds (SRTs) for the right ear revealed a slight loss in the ability to receive speech, whereas results for the left ear revealed a moderate to severe loss in the ability to receive speech. The SRTs were considered to be elevated in the right ear and in good agreement with pure-tone average in the left ear. Word recognition scores (WRSs) for the right ear revealed slight difficulty in the ability to recognize speech in the right ear and



AUDIOLOGICAL RECORD

TEST DATE
DOB SEX
AUDIOLOGIST
AUDIOMETER
EARPHONE

NAME

Effective Masking Level in dB

RE in	50	45 65	40 50	40 40	55 55	60 55	60	60			AC/BC
LE											AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	13	26		80%	66		
LT	57	56		28%	92 56		

Word Recognition List NU-6 1-A R L R L

RELIABILITY: Fair
VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.4	1.8

Static Admittance (mL): Rt 0.5 Lt 1.2
Peak Pressure (daPa): Rt 5 Lt -65
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI (x)	RT	RT	95	90				
	LT	LT	85	85				

COMMENTS:

Fig. 4.1 Audiological examination.

very poor ability to recognize speech in the left ear. Test reliability was judged to be fair due to the elevated SRT in the right ear and the presence of normal ipsilateral acoustic reflex thresholds (ARTs) at 500 to 1000 Hz for the right and left ears and the presence of type A tympanograms bilaterally in spite of a mixed moderate to moderately severe mixed loss in the left ear. A pure-tone Stenger test was negative at 500 and 1000 Hz.

4.3 Questions to the Reader

1. Based on the initial results of the audiological evaluation, what is suspected?
2. What further testing should be recommended?
3. How would you counsel this patient?

4.4 Discussion of Questions to the Reader

1. Based on the initial results of the audiological evaluation, what is suspected?
   Initial audiological results indicated an asymmetrical hearing loss, with the right ear revealing better hearing than the left ear. Due to the discrepancies in test results (i.e., lack of agreement between the SRT and pure-tone average in the right ear and lack of agreement between the ipsilateral ARTs in the left ear compared with the pure-tone thresholds for the left ear), a functional hearing loss is suspected. In addition, the audiogram revealed the presence of a mixed hearing loss in the left ear, but the tympanogram indicated normal middle ear function.
2. What further testing should be recommended?
   Several test options are possible. Otoacoustic emission (OAE) testing could be recommended, however, OAEs may not be measurable in the left ear due to the degree of hearing loss and the presence of the conductive component. Another option would be to evaluate the patient using nonbehavioral test methods, such as an auditory steady-state response (ASSR) test and/or an auditory brainstem response (ABR) test.
3. How should the patient be counseled at this time?
   If a functional hearing loss is suspected, the patient should be reinstructed to ensure that test procedures were understood. If repeat test results are still not in agreement, despite reinstructing the patient, tests that do not rely on behavioral responses should be recommended.

4.5 Diagnosis and Recommended Treatment

The patient was referred for ABR and ASSR tests due to the fair test reliability and asymmetrical hearing loss obtained during the initial evaluation.

4.6 Outcome

ASSR test results indicated hearing to be within normal limits in the right ear and a mild to moderate hearing loss in the left ear at 250 to 2000 Hz. ABR waveforms were established with a stimulus rate of 27.79 clicks per second, rarefaction polarity, and an amplifier gain of 240,000. Wave V peaks were identified at 40 dB nHL bilaterally, indicating no more than a mild hearing loss in either ear between 2000 and 4000 Hz. The patient was counseled regarding the results and that a functional hearing loss was suspected. The patient was referred for a repeat audiogram.

4.7 Key Points

1. Audiological test results should be compared for consistency.
2. If test results are not in agreement the patient should be reinstructed and retested.
3. Despite reinstruction, some patients can still present with test results that are not in agreement.
4. Electrophysiological testing can be used to estimate hearing sensitivity in cases where functional hearing is suspected.

Suggested Reading

Austen S, Lynch C. Non-organic hearing loss redefined: understanding, categorizing and managing non-organic behaviour. Int J Audiol 2004; 43: 449–457

McPherson B, McMahon K, Wilson W, Copland D. "I know you can hear me": neural correlates of feigned hearing loss. Hum Brain Mapp 2012; 33: 1964–1972

Monro DA, Martin FN. Effects of sophistication on four tests for nonorganic hearing loss. J Speech Hear Disord 1977; 42: 528–534

5 Auditory and Vestibular Neuropathy in an Adult Patient

Sarah L Grantham and Devin L McCaslin

JD is a 32-year-old female seen for an audiological evaluation in conjunction with a neurotology consultation due to concerns regarding sudden onset of bilateral hearing loss and tinnitus.

5.1 Clinical History and Description

JD is a 32-year-old female referred to the audiology clinic reporting a 5-month history of sudden bilateral hearing loss and tinnitus. Additional symptoms included dizziness, imbalance, blurred vision, and intermittent tingling and numbness of her face, tongue, hands, and feet. JD reported experiencing an "emotional breakdown" during a stressful life event with subsequent onset of all of her current medical symptoms. Prior to this incident, JD reported having normal hearing bilaterally and balance; no history of noise exposure, ear surgeries, or trauma; and no known family history of hearing loss. JD's medical history was also significant for anxiety, depression, and smoking one pack of cigarettes per day.

5.2 Audiological Testing

The initial audiological examination (□ Fig. 5.1a) was obtained with poor reliability and revealed a bilateral asymmetrical sensorineural hearing loss (SNHL). Results for the right ear revealed a moderate to severe SNHL at 250 Hz rising to a slight hearing loss at 8000 Hz. The speech recognition threshold (SRT) was 35 dB HL indicating a mild loss in the ability to receive speech. This was not in agreement with the moderately severe hearing loss exhibited by the pure-tone average (PTA) of 62 dB HL. The word recognition score (WRS) using a full-list of the Northwestern University Auditory Test No. 6 (NU-6) word lists with a recorded male talker at a presentation level of 105 dB HL revealed a WRS of 12% This revealed very poor ability to recognize speech and was an unexpected finding given the magnitude and configuration of the hearing loss. Results for the left ear revealed a moderate SNHL at 250 Hz rising to within normal limits at 8000 Hz. The (SRT) was 30 dB HL, indicating a mild loss in the ability to receive speech. This was not in agreement with the moderate hearing loss exhibited by the PTA of 45 dB HL. The (WRS) using a full-list of the NU-6 word lists with a recorded male talker at a presentation level of 90 dB HL revealed a WRS of 52% This revealed a poor ability to recognize speech and was also an unexpected finding given the magnitude and configuration of the hearing loss. Immittance audiometry was completed, and the resulting tympanograms were consistent with normal middle ear function. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were absent bilaterally. Distortion-product otoacoustic emissions (DPOAEs) were present at 750 to 8000 Hz bilaterally (□ Fig. 5.2).

5.3 Questions to the Reader

1. Given JD's medical history and initial audiological findings, what impressions can be drawn with respect to potential cause(s) of her hearing loss?
2. At this point, would further diagnostic testing be recommended, and if so, what additional tests would be recommended?
3. What indications from the initial audiological evaluation would warrant auditory brainstem response (ABR) testing?

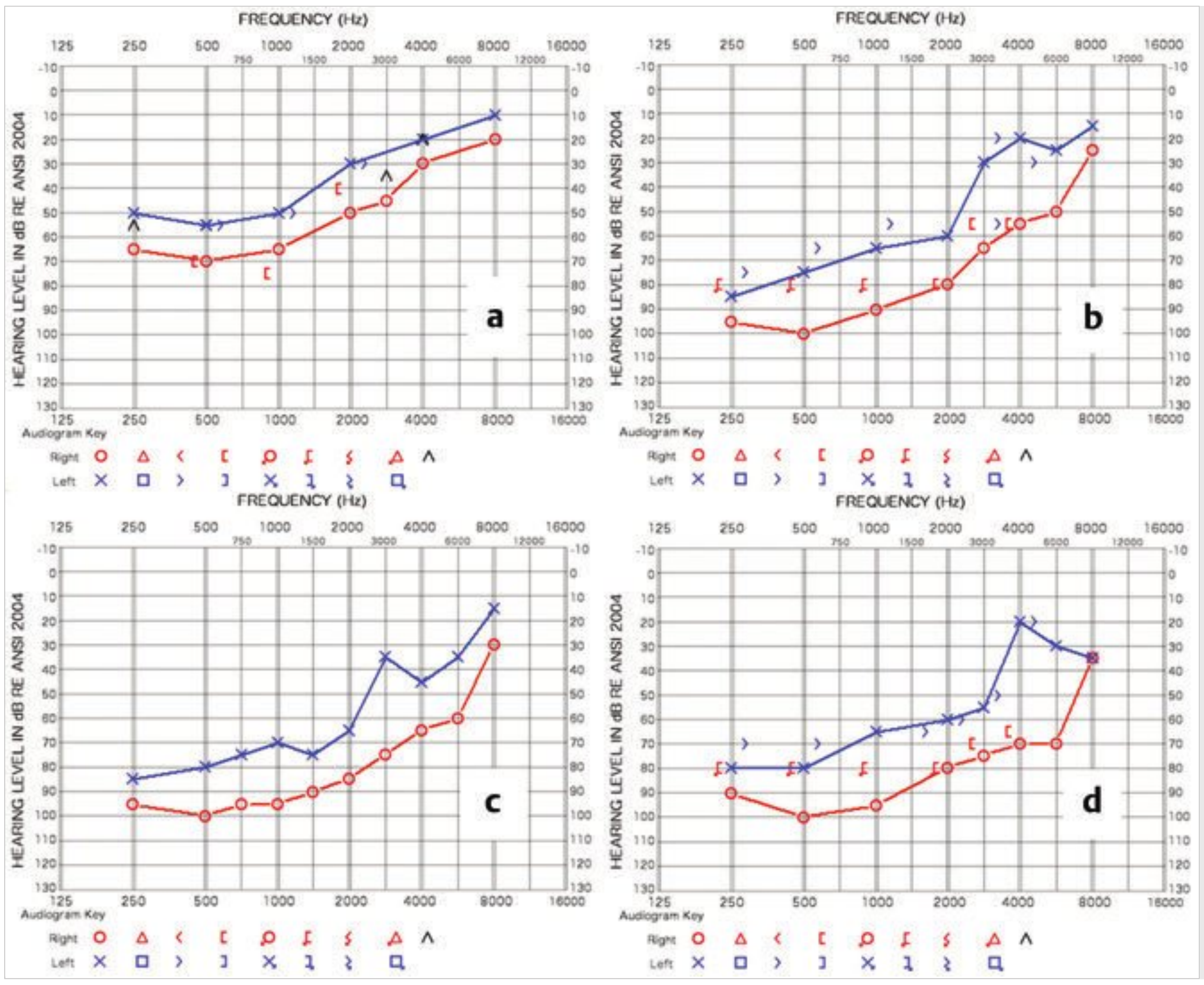


Fig. 5.1 Audiogram revealing the fluctuant nature of JD's hearing loss. (a) Initial audiogram. (b) Three months following the initial audiogram. (c). Five months following the initial audiogram. (d) Fourteen months following the initial audiogram.

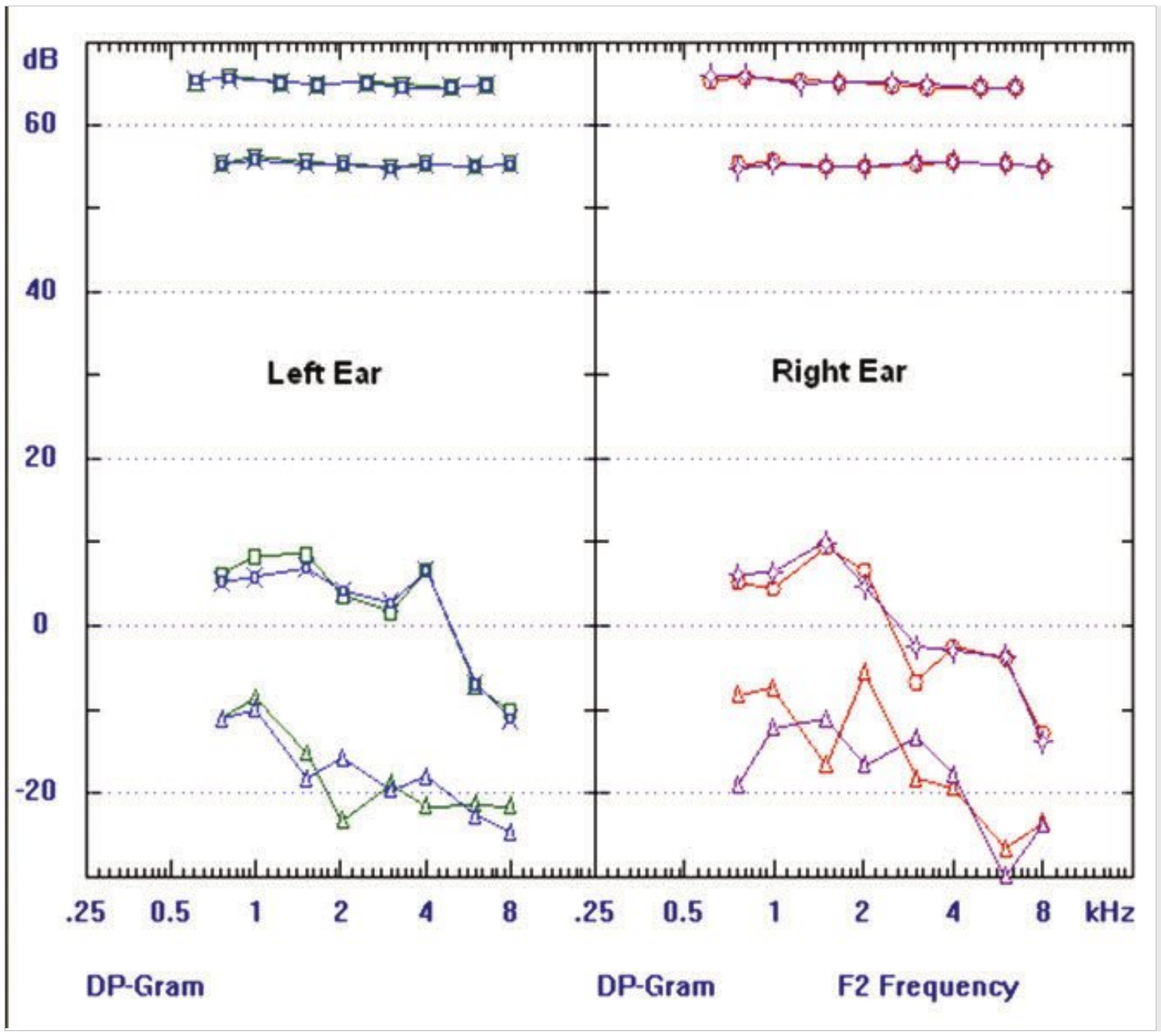


Fig. 5.2 Distortion-product otoacoustic emissions present for all test frequencies, bilaterally.

5.4 Discussion of Questions to the Reader

1. Given JD's medical history and initial audiological findings, what impressions can be drawn with respect to potential cause(s) of her hearing loss?
   Given the poor reliability, disagreement between PTAs and SRTs, disproportionately poor WRSs, and present DPOAEs, the potential for JD's hearing loss to be nonorganic in nature would be considered. The absence of ARTs to ipsilateral and contralateral stimulation, however, suggests evidence of neural hearing loss. Whereas DPOAEs assess the functional integrity of the cochlear outer hair cells (OHCs), ARTs assess the neural integrity of the auditory eighth cranial nerve. Collectively, this pattern of findings (i.e., absent ARTs with the presence of DPOAEs) is commonly manifested in patients with auditory neuropathy spectrum disorder (ANSD) and should be considered in the differential diagnosis.
2. At this point, would further diagnostic testing be recommended, and if so, what additional tests would be recommended?
   ABR testing could serve to confirm or deny the presence of nonorganic hearing loss (NOHL) versus ANSD. Additionally, given JD's report of dizziness and imbalance, comprehensive vestibular function testing (VFT) to include electrovideonystagmography (ENG/VNG), sinusoidal harmonic acceleration (SHA), and ocular and cervical vestibular-evoked myogenic potentials (oVEMPs and cVEMPs) would also be indicated.
3. What indications from the initial audiological evaluation would warrant ABR testing?
   JD demonstrated DPOAEs that were present. JD's DPOAEs should have been absent given the magnitude of hearing loss reported at the initial audiological evaluation. The absence of ARTs to contralateral and ipsilateral stimulation at all test frequencies with present DPOAEs is an important indicator for ABR testing.

5.5 Additional Testing

Due to the inconsistent audiological results, ABR testing was recommended to obtain a more objective measure of JD's underlying hearing sensitivity. ABR testing was completed with a high-frequency click stimulus. At 85 dB nHL, no repeatable neural responses were observed bilaterally using condensation, rarefaction, and alternating polarity. A cochlear microphonic (CM), however, that reversed with a change in stimulus polarity, was noted bilaterally (Fig. 5.3). The CM disappeared when the stimulus tubing was clamped, thus ruling out stimulus artifact. JD's ABR results were consistent with ANSD, and this led to a referral for an evaluation of candidacy for a cochlear implant (CI).

Additionally, due to JD's reports of dizziness and imbalance, comprehensive vestibular function testing (VFT) was also ordered. The VFT included videonystagmography (VNG), sinusoidal harmonic acceleration (SHA), and cVEMP and oVEMP examinations. The VNG revealed normal ocular motility and positional/positioning subtests, with bilaterally abnormal caloric responses. Specifically, the total caloric response (i.e., 13°/s where <30°/s is abnormal) was significantly reduced. SHA results revealed abnormally reduced vestibulo-ocular reflex

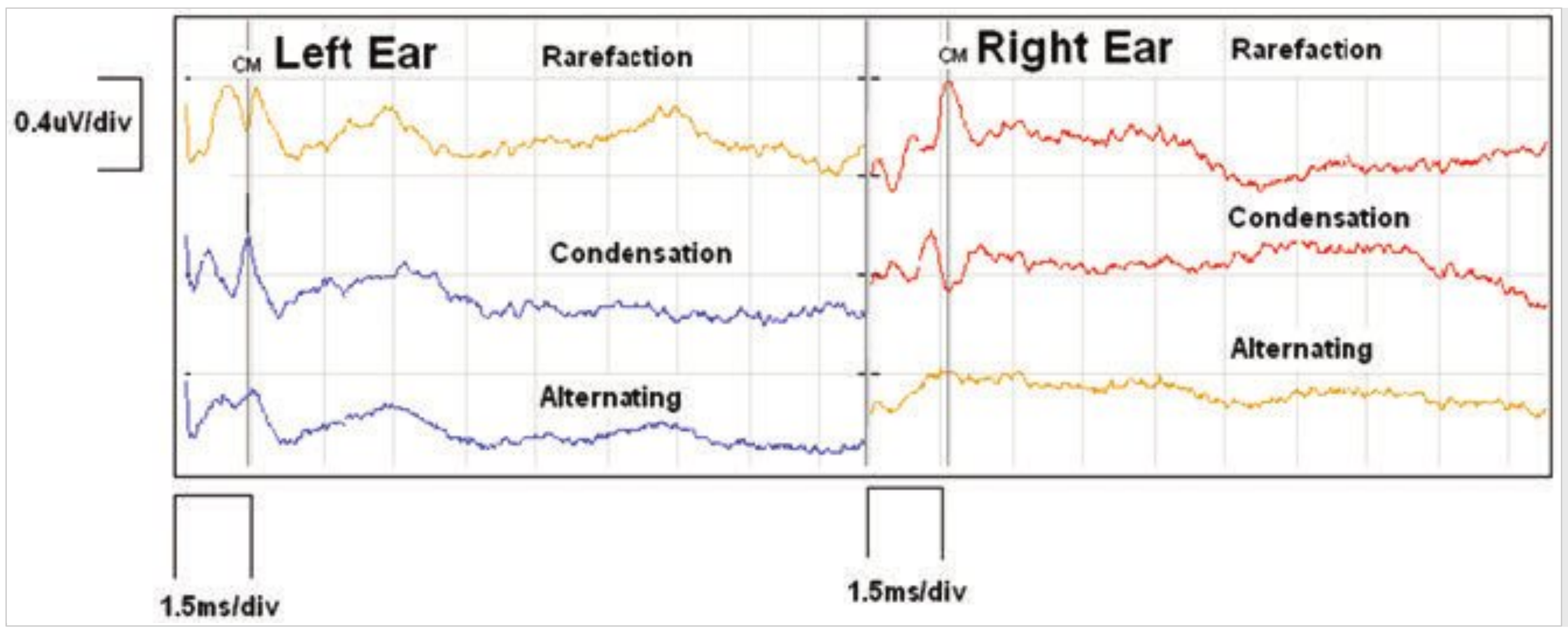


Fig. 5.3 Averaged auditory brainstem response waveforms obtained using rarefaction, condensation, and alternating polarity clicks.

(VOR) gain across multiple frequencies. The cVEMPs and oVEMPs were absent bilaterally.

5.6 Additional Questions to the Reader

1. Based on the audiological and vestibular test results, what conclusions can be made with regard to the cause of JD's hearing loss, dizziness, and imbalance?
2. What types of treatment, intervention, and/or rehabilitation options would be recommended based on the finding of impaired vestibular function?
3. What genetic conditions should be considered in the differential diagnosis?
4. What are the implications of vestibular neuropathy?

5.7 Discussion of Additional Questions to the Reader

1. Based on the audiological and vestibular test results, what conclusions can be made with regard to the cause of JD's hearing loss, dizziness, and imbalance?
   Imbalance and unsteadiness can occur in patients with auditory neuropathy when a generalized peripheral neuropathy is present. It is known that patients with auditory neuropathy can also present with abnormal vestibular test findings (e.g., caloric and VEMP), the results of which are attributed to neuropathy of the vestibular nerve. Reports have shown that patients with auditory neuropathy can develop impairment of the vestibular nerve. Unfortunately, in many audiology centers, routine vestibular function testing on patients with confirmed neuropathic disorders is not typically provided. The fluctuating audiometric thresholds (▯ Fig. 5.1b–d) combined with the additional findings from the auditory and vestibular electrophysiological tests suggest that JD probably has auditory and vestibular neuropathy.
2. What types of treatment, intervention, and/or rehabilitation options would a clinician recommend based on the finding of impaired vestibular function?
   The identification of vestibular dysfunction suggests JD should be referred to a healthcare provider who specializes in vestibular rehabilitation. That is, JD may benefit from a therapeutic approach directed at improving vestibular function. JD, however, should initially be evaluated by a neurologist for any additional generalized neuropathies that may be impairing gait and standing balance. A referral to a genetic counselor would also be recommended in order to rule out potential hereditary causes of JD's auditory and vestibular neuropathy.
3. What genetic conditions should be considered in the differential diagnosis?
   Charcot–Marie–Tooth disease (CMT)—Named after the three scientists who first described the condition, CMT is now classified within a group of interrelated neurodegenerative disorders known as hereditary motor and sensory neuropathy (HSMN). These disorders are caused by axonal loss or demyelination and cause muscular atrophy, sensory loss, and diffuse peripheral neuropathies.
   Friedreich ataxia—An autosomal recessive inherited degenerative neuromuscular disorder caused by reduced production of the protein frataxin. Degeneration of the spinal cord and peripheral nerves may result in visual impairments, hearing loss, speech and swallowing disorders, and ataxic gait.
   Otoferlin (OTOF)—Mutation(s) of the OTOF gene affect production of the protein otoferlin, causing nonsyndromic deafness in the form of auditory neuropathy.
   Pejvakin (PJVK)—PJVK is a protein found within the afferent auditory pathway. Genetic mutations that affect the encoding sequence of PJVK have been shown to result in autosomal recessive auditory neuropathy.
   Autosomal dominant mutations (AUNA1)—The AUNA1 gene has been found to cause a rare form of progressive autosomal dominant auditory neuropathy.
4. What are the implications of vestibular neuropathy?
   Patients with vestibular neuropathies generally do not report vertigo as typically experienced by patients who abruptly lose peripheral vestibular function (e.g., vestibular neuritis). Rather, patients with vestibular neuropathies complain of gradual unsteadiness and generalized imbalance. These symptoms are most likely the result of the slowly progressive and bilateral nature of vestibular neuropathy. In this regard, patients with vestibular neuropathy may also report oscillopsia (i.e., a visual perception that stationary objects are moving) and decreased balance in low-light situations (e.g., getting up at night). Balance dysfunction in patients with vestibular neuropathy may also be enhanced when there is a concomitant generalized peripheral neuropathy that reduces lower extremity sensation and impairs proprioception.

5.8 Diagnosis and Recommended Treatment

The results from a comprehensive audiological evaluation and follow-up vestibular function testing were consistent with bilateral auditory and vestibular neuropathy. JD was referred for an evaluation for candidacy for a CI and to investigate additional potential aural rehabilitation options. Vestibular rehabilitative therapy (VRT) was recommended to address JD's continuing reports of imbalance. In addition, referrals to neurology and genetic counseling were recommended to further investigate the origin of JD's disease process.

5.9 Outcome

JD was found to be a good candidate for bilateral CIs. She decided to receive a CI for her right ear. Unfortunately, initial insurance coverage of this procedure was denied. The appeal process lasted 18 months. In the end, JD received a CI for her right ear. JD is performing well with her CI, responding to auditory stimuli and reporting that she is able to hear new sounds every day (e.g., birds, flip-flops, car horns, etc.). JD reports completing aural rehabilitation exercises 30 minutes daily and wears her CI full time.

JD's neurological examination remains inconclusive. Magnetic resonance imaging (MRI) and computed tomographic (CT) scans of the brain and spinal cord were normal. Bloodwork for rheumatoid arthritis, thyroid disease, autoimmune disease, and syphilis were negative. Although JD was found to have an intact sense of vibration in her lower extremities, JD continues to report poor balance and frequent (i.e., weekly) falls. These reports reinforce the need for VRT as a therapeutic intervention strategy for treatment of JD's underlying bilateral peripheral vestibular neuropathy.

5.10 Key Points

1. ANSD can be identified at any age. In adults, hearing thresholds can be stable, fluctuate, or become progressively poorer. This fluctuation depends on the underlying cause of the ANSD.
2. In patients with a diagnostic pattern of auditory findings suggesting ANSD, the vestibular system should also be evaluated. The bithermal caloric test and oVEMP and cVEMP examinations can evaluate the integrity of the two divisions (i.e., inferior and superior) of the vestibular nerve.
3. In many cases, patients with ANSD often have accompanying peripheral neuropathies. Although these patients may complain of generalized imbalance, it is the clinician's role to determine whether the unsteadiness is attributed to vestibular dysfunction (i.e., impairment of the vestibular nerve), chronic peripheral neuropathy, or both.

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6 You Really Should See a Doctor about That

Lauren Harvey and Wayne J. Wilson

A 55-year-old male purchased two behind-the-ear (BTE) hearing aids online and has presented to an audiologist to have his hearing aids fitted.

6.1 Clinical History and Description

AB is a 55-year-old male who over the past 5 years has found it increasingly difficult to hear in noisy listening environments. Being an electrical engineer, AB felt confident enough to conduct his own extensive online research on hearing loss and hearing aids. AB concluded that his hearing loss was the result of his advancing age, that he would benefit from hearing aids, and that the most financially efficient option would be to purchase the hearing aids online and have them fitted by an audiologist.

On presentation to the audiologist, AB reported his hearing had slowly deteriorated over the past 10 years, being particularly noticeable over the past 5 years. AB now feels his hearing affects his ability to communicate with his family and clients, particularly when there is background noise, if there are multiple talkers, or if AB is more than a few meters away from the speaker. AB reported no difference in hearing between his ears; no difficulties hearing the television, although he admitted to occasionally needing to increase the volume; and no difficulties hearing on the telephone, which he typically holds to his left ear so he can write with his right hand. When asked about tinnitus, AB reported hearing a constant, low-level "cicada/cricket-like" sound bilaterally for as long as he can remember, and although this sound had never bothered him it had become worse in his right ear over the past few years. AB did not report any vertigo or general dizziness. When asked about noise exposure, AB reported that he worked as an electrical engineer, initially in the public sector, and for the past 10 years in the private sector where he manages his own engineering practice. While sometimes exposed to loud noise, AB reported always wearing appropriate hearing protection after not always having done so early in his career. AB reported no history of hearing loss in his immediate family, stating that his parents had good hearing into their advancing years. Finally, AB reported he was in good health and had not experienced any major medical events since having his appendix removed when he was in primary school.

6.2 Audiological Testing

Fig. 6.1 reports the results of AB's audiological examination. Pure-tone testing revealed a bilateral asymmetric sensorineural hearing loss (SNHL) of a mild to moderately severe degree above 1000 Hz in the right ear and a slight to moderate degree above 1000 Hz in the left ear. Because this testing was completed in Australia, AB's word recognition scores (WRSs) were obtained using the National Acoustics Laboratories' Arthur Boothroyd (NAL-AB) word lists and its recommended protocols. The NAL-AB wordlists contain 15 lists of 10 monosyllabic words. The patient's response to each word is scored phonemically (allowing for 30 phonemes per list) such that correctly repeating three, two, one, or none of the phonemes in each word elicits a score of 10%, 7%, 3%, or 0% for that word, respectively. A performance-intensity (PI) function is obtained for each ear by presenting the first word list at the expected half-maximum level (HML), the level at which the patient is predicted to score 50%, and subsequent lists at levels 15 dB higher than the previous list until a maximum score is measured. If not already revealed using the first word list, a score is also obtained at a signal level below HML. Speech noise is applied to the nontest ear according to the following formula: masking level in the nontest ear = presentation level in the test ear (–) 40 dB + the air–bone gap in the nontest ear + audiometer conversion factor. Once the PI functions are plotted, the speech recognition threshold (SRT) is estimated by extrapolating the presentation level required to reach the 50% score on the PI function. AB's SRTs were in agreement with the pure-tone average and revealed a mild loss in the ability to receive speech in the right ear and a slight loss in the ability to receive speech in the left ear. AB's WRSs revealed slight difficulty in recognizing speech in his right ear and normal ability to recognize speech in his left ear. As a side note, AB's WRSs improved with increased presentation levels to a maximum WRS of 87% in the right ear and 100% in the left ear with no significant rollover at the 90 dB HL presentation.

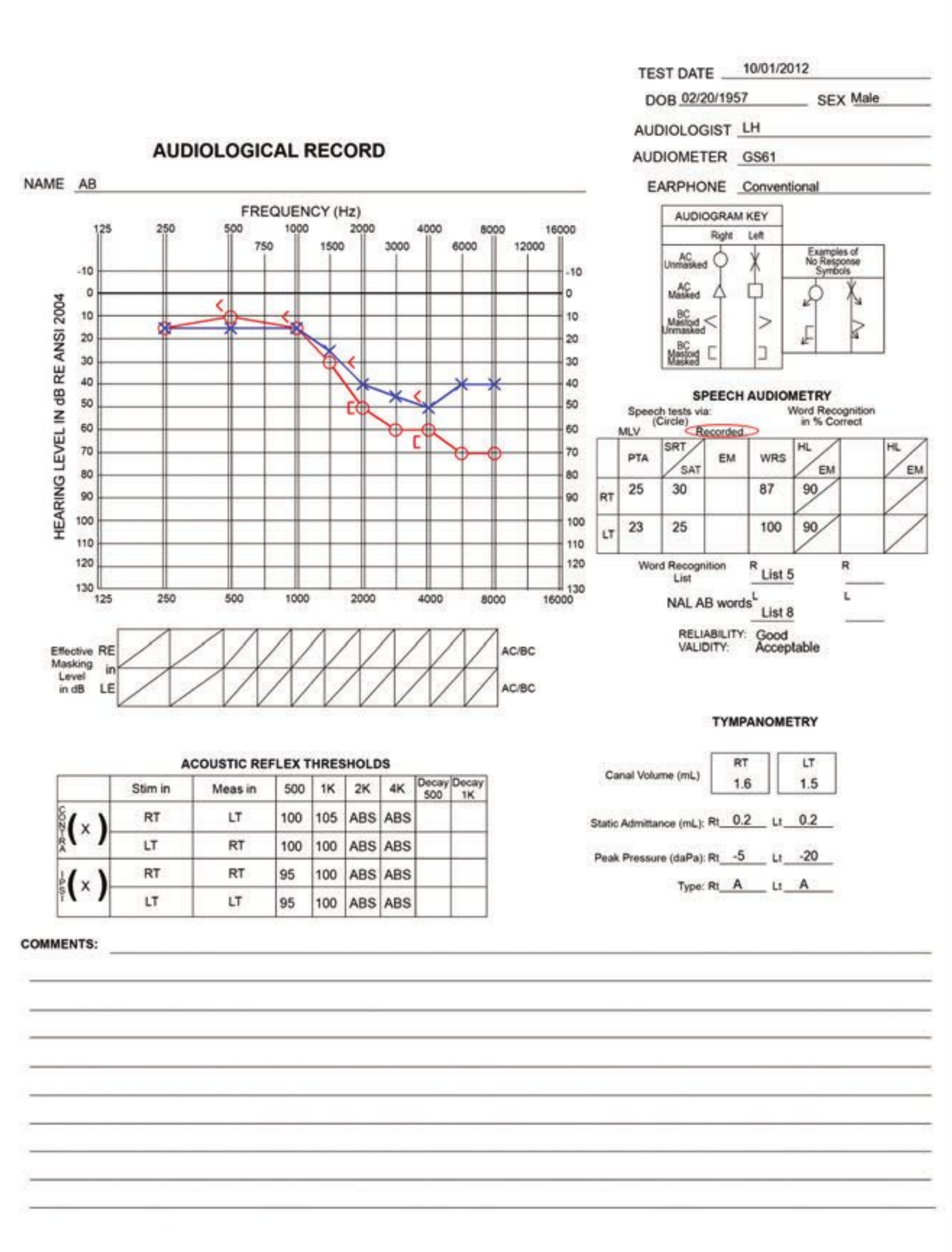
Tympanometry revealed that middle ear pressure (daPa), ear canal volume (mL), and static admittance (mL) were within normal range bilaterally, although, the static admittance of 0.2 mL was at the lower end of the normal range bilaterally. The contralateral and ipsilateral acoustic reflex thresholds (ARTs) were present at levels from 95 dB HL to 105 dB HL at 500 Hz and 1000 Hz, but absent at 2000 Hz and 4000 Hz bilaterally. These ARTs are consistent with the audiometric and tympanometric results.

6.3 Question to the Reader

1. Should the audiologist proceed with AB's hearing aid fitting?

6.4 Discussion of the Question to the Reader

1. Should the audiologist proceed with AB's hearing aid fitting?
   Several factors within AB's case history and audiological examination suggest he is a good candidate for amplification. AB's case history shows he is highly motivated with a positive attitude toward hearing aids because he already conducted his own research and purchased his own hearing aids online. AB's audiological results reveal he has bilateral high-frequency SNHL that falls within a range that can be aided,



AUDIOLOGICAL RECORD

NAME AB

TEST DATE 10/01/2012
DOB 02/20/1957 SEX Male
AUDIOLOGIST LH
AUDIOMETER GS61
EARPHONE Conventional

Effective Masking Level in dB	RE in											AC/BC
	LE											AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	25	30		87	90		
LT	23	25		100	90		

Word Recognition List NAL AB words R List 5 L List 8 R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.6	1.5

Static Admittance (mL): Rt 0.2 Lt 0.2
Peak Pressure (daPa): Rt -5 Lt -20
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	100	105	ABS	ABS		
	LT	RT	100	100	ABS	ABS		
IPSI (X)	RT	RT	95	100	ABS	ABS		
	LT	LT	95	100	ABS	ABS		

COMMENTS:

Fig. 6.1 AB's audiological examination.

and his WRSs were reported to improve with increased presentation level bilaterally without any sign of rollover (i.e., decreased WRS as the presentation level is increased).

Although the foregoing factors are encouraging, closer inspection of AB's case history and audiological results reveal some areas of concern. Despite AB's substantial efforts to research hearing and hearing loss, his conclusion that his hearing loss is solely the result of his advancing age (presbycusis) is premature for at least two reasons. First, AB's longstanding bilateral tinnitus has become worse in his right ear over the past few years. Second is AB's asymmetric SNHL despite AB reporting no difference in hearing between his ears in the case history. These two concerns highlight the significant weighting given by audiologists to asymmetric findings when deciding if further diagnostic examination is indicated. This weighting is based on the assumption that otologically normal patients (i.e., patients with normal health who are free of symptoms of ear disease and from obstructing cerumen in the ear canals and who have no history of exposure to noise) will have symmetric hearing. This assumption suggests that patients with asymmetric hearing could have an otologic abnormality.

Quantifying the term asymmetric can be challenging, however, with several criteria published in the literature. Some of these criteria emphasize asymmetries at single frequencies (e.g., > 15 dB at any frequency), others emphasize asymmetries at multiple frequencies (e.g., > 15 dB at two or more frequencies, or > 15 or 20 dB at two adjacent frequencies), while still others emphasize asymmetries in a pure-tone average (PTA) (e.g., > 15 dB in PTAs calculated from all octave frequencies or various subsets of the audiometric frequencies). Finally, many of these criteria also include other factors such as sudden SNHL, unilateral tinnitus, vertigo, asymmetric WRSs, and others. In AB's case, his audiological results met several, but not all, published criteria for asymmetric hearing loss. For example, AB's right ear had greater tinnitus and poorer hearing thresholds by > 15 dB at one frequency and > 20 dB at two adjacent frequencies. Also, his right ear had the poorer WRS.

In light of AB's case history and audiological results, the audiologist recommended that AB defer his hearing aid fitting until AB had undergone an otologic examination to rule out any medical contraindications for fitting his hearing aids and to determine if his hearing loss required nonaudiological management. AB objected to this recommendation, stating that the audiologist should stop “wasting his time and money” and fit him with his hearing aids immediately. The audiologist then counseled AB on the significance of his asymmetric audiological findings and the need to have this investigated. The audiologist also informed AB that she was not willing to proceed with his hearing aid fitting until AB had completed the otologic examination and that she could refer AB to another audiologist if he wished to seek a second opinion. AB gave this some thought before finally accepting the audiologist’s recommendation.

6.5 Additional Testing

After examining AB’s ears and reporting “no abnormalities detected,” AB’s otologist referred him to a radiologist for a magnetic resonance imaging (MRI) scan. The radiologist reported that on T2-weighted images taken after an intravenous application of gadolinium (a contrast medium used to improve the visibility of internal body structures on MRI scans), the right ear revealed a 5 mm nodular mass in the right internal auditory canal with some subtle widening of the right porus acusticus internus (the opening to the internal auditory canal). The left ear revealed a normal internal auditory canal and seventh/eighth cranial nerve complex (the nerve complex formed by the seventh cranial nerve, the facial nerve, and the eighth cranial nerve, the vestibulocochlear nerve), which passes through the internal auditory canal.

6.6 Diagnosis and Recommended Treatment

The otologist informed AB that he believed the mass in his right internal auditory canal was most likely an acoustic neuroma. An acoustic neuroma is a benign (i.e., does not spread to other parts of the body) tumor that involves the eighth cranial nerve. It is the most common tumor in the internal acoustic canal and cerebellopontine angle (CPA), the angle created by the cerebellum and the pons in the brainstem; it is slowly growing; it can compress or destroy nearby structures such as the seventh and eighth cranial nerves and it can cause symptoms such as hearing loss, tinnitus, or vertigo. Large acoustic neuromas can also compress the brainstem, which can affect life-sustaining functions such as the maintenance of heartbeat and breathing. Because acoustic neuromas typically arise from the Schwann cells (the fatty cells that wrap around nerve axons to increase nerve impulse velocity) of the vestibular portion of the eighth cranial nerve, they are more accurately called vestibular schwannomas.

The otologist also informed AB of the treatment options for acoustic neuromas. The first was to monitor, which involves regularly scheduled MRI scans with no action taken as long as the tumor does not grow. The second option was microsurgery, which involves anesthesia, an incision in the skull, and the physical removal of the tumor from the eighth cranial nerve. The third option was stereotactic radiosurgery, which involves the delivery of a focused and precise dose of radiation to the tumor either as a single dose or over multiple sessions. After discussing the advantages and disadvantages of each option with his otologist, AB elected to undergo stereotactic radiosurgery.

After completing the additional testing, AB returned to the audiologist to request that he now be fitted with his hearing aids.

6.7 Additional Question to the Reader

1. Should the audiologist now proceed with AB’s hearing aid fitting?

6.8 Discussion of the Additional Question to the Reader

1. Should the audiologist now proceed with AB’s hearing aid fitting?

   The factors that initially suggested AB was a good candidate for hearing aids were still in place after he had been diagnosed with an acoustic neuroma in his right ear and had decided to undergo stereotactic radiosurgery. The factors that were of significant concern and that led the audiologist to refer AB for further otologic examination had now been explained and were being appropriately managed. Also, AB’s otologist had been able to rule out any medical contraindications for fitting AB with his hearing aids. With these factors in place, AB’s audiologist decided to fit AB with his hearing aids and to monitor AB’s hearing and hearing aids on a quarterly basis. The audiologist also counseled AB on the importance of this monitoring for his hearing and for any adjustments needed to his hearing aids should his hearing change as a result of his acoustic neuroma and/or radiosurgery.

   In reviewing this case, AB’s audiologist noted she could have begun AB’s hearing aid fitting after the initial audiological testing, but before the further otologic (and eventually radiological) examination. This is true because AB had already purchased his hearing aids prior to seeing the audiologist, and the hearing aids could have been adjusted as the otologic diagnostic results became available. In light of the final diagnosis, however, the audiologist was satisfied that delaying the fitting pending further otologic assessment had not only allowed AB to concentrate on identifying one of the true causes of his hearing loss but had also allowed the audiologist to use the final diagnosis to provide AB with more complete audiological counseling. This was especially true with regard to AB’s acceptance of his hearing loss and its cause(s), what he should expect from his hearing aids, and the importance of monitoring his hearing and hearing aid performance in the future.

6.9 Outcome

The audiologist successfully fitted AB with his previously purchased hearing aids and began to monitor AB’s progress as he underwent stereotactic radiosurgery for his acoustic neuroma.

6.10 Key Points

1. Factors such as the public's increasing ability to directly access hearing aids does not diminish audiologists' duty to appropriately assess and manage their patients.
2. Asymmetric audiometric thresholds carry significant weight when determining if further otologic examination is indicated, although quantifying the degree of asymmetry required to trigger such further examination remains a challenge.
3. Although some contraindications to hearing aid fitting are absolute, others (such as the presence of an acoustic neuroma) can be conditional.

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7 Acoustic Neuroma

Nancy Caryn McLellan and Ross J. Roeser

BP is a 31-year-old male who was referred because he reported hearing loss in his left ear, which was later confirmed to be the result of an acoustic neuroma (AN).

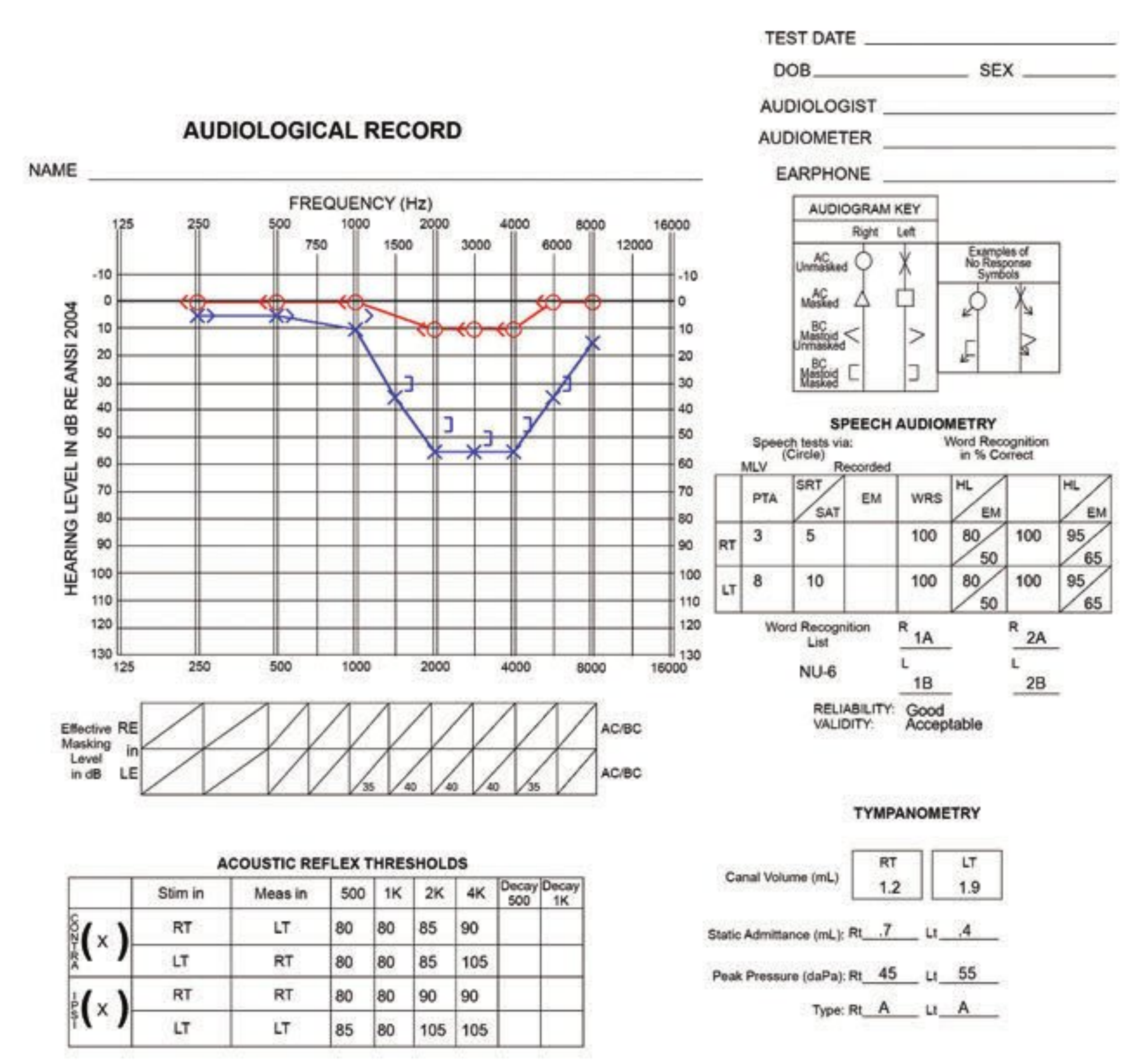
7.1 Clinical History and Description

BP, a 31-year-old male, reported progressive, fluctuating hearing loss in the left ear with aural fullness. He indicated the symptoms were present for approximately a year and a half prior to his appointment for an audiological examination. Two months prior to his audiology appointment, he consulted an urgent care physician who treated him with steroids. BP experienced no improvement in his symptoms and subsequently discontinued the use of the steroids. Six weeks prior to the audiology appointment, BP experienced a sudden relief in aural fullness, but the hearing loss in his left ear remained persistent and was then accompanied by unilateral tinnitus. At his appointment, BP denied otalgia, otorrhea, aural fullness, and dizziness. He reported a 10-year history of military service and recreational noise exposure involving using rifles held on the right shoulder. Despite having an audiometric configuration consistent with possible noise-induced hearing loss and no other findings consistent with retrocochlear involvement, radiological studies confirmed BP had a large AN pressing against the left middle cerebellar peduncle.

7.2 Audiological Testing

BP was evaluated with behavioral and electrophysiological audiological procedures. As reported in Fig. 7.1, pure-tone audiometry revealed hearing to be within normal limits in the right ear at 250 to 8000 Hz. Results for the left ear revealed a mild to moderate mid- to high-frequency sensorineural hearing loss at 1500 to 6000 Hz with improvement to within normal limits at 8000 Hz. It should be noted that pure-tone thresholds for the left ear would be expected for a patient with a history of noise exposure who used a rifle held on the right shoulder. Speech recognition thresholds (SRTs) revealed normal ability to recognize speech bilaterally and are in agreement with the three-frequency pure-tone average for the right ear. A two-frequency average at 500 and 1000 Hz was used for the left ear. Word recognition scores (WRS) using recorded versions of the Northwestern University Auditory Test No. 6 (NU-6) word lists revealed normal ability to recognize speech bilaterally. In addition, WRSs did not decrease as the presentation level was increased from 80 to 95 dB HL, indicating no significant rollover.

Tympanometry revealed slight positive pressure for the right (+45 daPa) and left (+55 daPa) ears but was considered to be within the normal range. Ear canal volume (mL) and static admittance (mL) were also within the normal range. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were normal, although the ART at 4000 Hz was elevated in the stimulus



TEST DATE
DOB
SEX
AUDIOLOGIST
AUDIOMETER
EARPHONE

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	3	5		100	80 / 50	100	95 / 65
LT	8	10		100	80 / 50	100	95 / 65

Word Recognition List NU-6 R 1A L 1B R 2A L 2B

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.9

Static Admittance (mL): Rt .7 Lt .4
Peak Pressure (daPa): Rt 45 Lt 55
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	80	80	85	90		
	LT	RT	80	80	85	105		
IPSI (X)	RT	RT	80	80	90	90		
	LT	LT	85	80	105	105		

Fig. 7.1 Audiological results for BP.

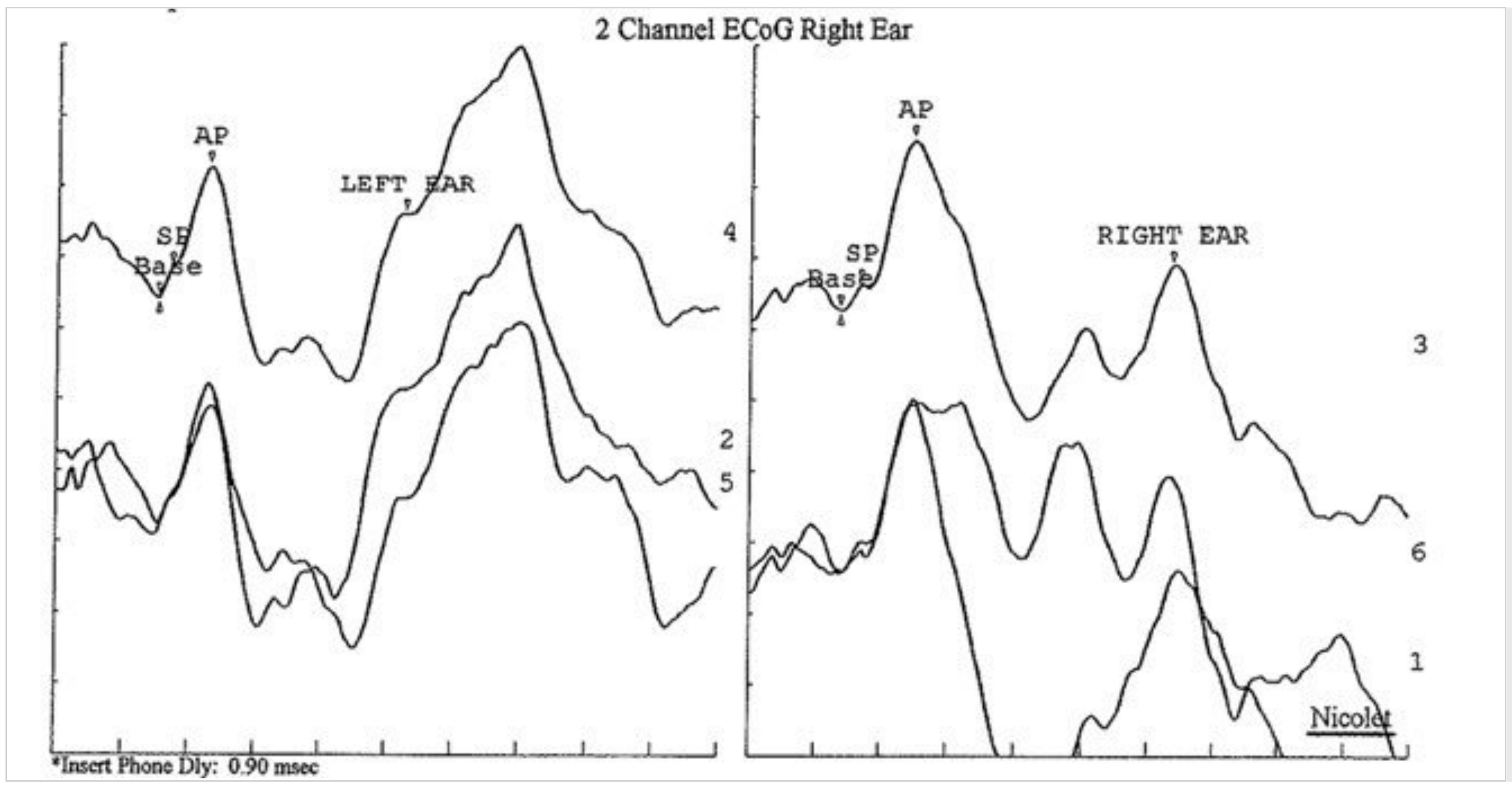


Fig. 7.2 Electrocochleography results for BP indicating normal findings bilaterally

left contralateral and ipsilateral conditions as well as at 2000 Hz in the ipsilateral stimulus left condition.

Due to the asymmetry revealed on the audiogram and the patient report of aural fullness and unilateral tinnitus, the patient was referred for electrocochleography (ECoG). As shown in Fig. 7.2, ECoG waveforms were robust with normal latencies and summating potential (SP)/action potential (AP) ratios. This finding ruled out the possible presence of excess fluid in the cochlea, which might be indicative of Ménière disease. Based on these data, referral to an otoneurologist was immediately made. At the subsequent appointment the physician ordered a magnetic resonance imaging (MRI) scan.

7.3 Questions to the Reader

1. Are there other terms used to describe an AN?
2. What is the role of an audiologist in diagnosing ANs, and when should an audiologist consider retrocochlear involvement as a possible etiology for the hearing loss?
3. The use of acoustic reflex decay testing has recently become questionable due to its potential to cause temporary or permanent threshold shift. Is the use of acoustic reflex decay warranted when an AN is suspected?
4. Should a vestibular evaluation be performed in the absence of vestibular complaints?

7.4 Discussion of Questions to the Reader

1. Are there other terms used to describe an AN?
   Yes, ANs are slow-growing, typically benign, tumors that originate in the Schwann cell lining of the vestibular branch of the eighth cranial nerve. The AN can cause hearing loss and vestibular symptoms as it grows because the enlarging mass begins to press against the auditory branch of the eighth cranial nerve in the internal auditory canal. Although these growths are commonly called acoustic neuromas due to the auditory symptoms they create, the term vestibular schwannoma is more accurate. The terms neurilemmoma and neurinoma have also been used to describe ANs.
2. What is the role of an audiologist in diagnosing ANs, and when should an audiologist consider retrocochlear involvement as a possible etiology for the hearing loss?
   Audiologists aid in the diagnosis of ANs by performing conventional and advanced diagnostic procedures that are considered functional tests of the auditory system. There are two types of assessments involved in AN diagnosis: structural and functional. Structural or anatomical assessments, such as MRI scans, are those that evaluate the anatomy of the patient's central nervous system. Only a physician is qualified and licensed to determine if further medical testing, including radiographic imaging, is necessary to rule out the presence of retrocochlear pathology.

   Functional (audiological) assessments such as the audiological or vestibular test battery are used to evaluate the capability of each auditory or vestibular structure to perform its task. Although the MRI scan is the gold standard for the detection of ANs and is required for definitive diagnosis, diagnostic audiological procedures provide unique and valuable information about the integrity and functional capacity of a patient's auditory system. Diagnostic audiological examinations are important in aiding physicians and surgeons to determine treatment options and follow-up care. In addition, diagnostic audiological procedures are needed to determine pre- and posttreatment audiological rehabilitation, including counseling and technological rehabilitation (hearing instruments, hearing assistive devices, and possibly auditory brainstem implants).

   The symptoms typically associated with AN include sudden or gradual unilateral hearing loss, aural fullness, tinnitus (typically unilateral), WRS score that is poorer than predicted from the pure-tone audiogram, dizziness, and/or facial numbness or weakness. Audiological tests for retrocochlear involvement should be considered whenever any of these symptoms are present. Among the procedures that should be considered are auditory brainstem response (ABR) testing, auditory steady-state response (ASSR) tests, threshold tone decay, and possibly acoustic reflex decay. WRS can also be used as a screening tool to assess the presence of rollover, which is a significant reduction in WRS performance once the Maxium Performance (PBmax) on the word recognition reached. A retrocochlear diagnosis is considered when the rollover index is 0.45 or greater (PBmax − PBmin(the poorest performance)/PBmax). A detailed discussion of each of these procedures can be found in Roeser et al.

3. The use of acoustic reflex decay testing has recently become questionable due to its potential to cause temporary or permanent threshold shift. Is the use of acoustic reflex decay warranted when an AN is suspected?
   The stimulus for the acoustic reflex decay test is a continuous pure-tone presented for 10 seconds at 10 dB sensation level (SL) about the ART at 500 and 1000 Hz for contralateral stimulation. Significant decay as a positive finding for retrocochlear pathology is defined as a decrease in maximum amplitude to less than half within 5 seconds. Although the acoustic reflex decay test has a moderately high sensitivity rate for diagnosing retrocochlear lesions, there is the risk of the test causing a temporary or permanent threshold shift due to the high presentation level required to administer the test. As a result, it has been suggested that acoustic reflex decay testing should not be used in diagnostic assessments. When other tests are suggestive of AN, however, the use of the acoustic reflex decay test is possible but should be used judiciously, with the presentation level prevented from exceeding 105 to 110 dB HL. Pure-tone thresholds and ABR testing have been shown to be more sensitive to the presence of ANs without the possibility of causing further damage to hearing.
4. Should a vestibular evaluation be performed even in the absence of vestibular complaints?
   Electronystagmography (ENG) or videonystagmography (VNG) and other vestibular function tests can be used to assess the function of the vestibular system. Although 50% of patients with ANs experience impaired balance, vestibular testing may not be routinely performed for patients with ANs. Performing this assessment before treatment is important because it can provide a baseline that can later be used to measure outcome success after treatment. In addition, ENG or VNG and other measures can be used to monitor progression of symptoms if the AN should grow.

7.5 Additional Testing

7.5.1 Audiological

For this case, the audiologist referred the patient to an otologist who referred the patient for an MRI scan following the comprehensive audiological examination and ECoG. Three additional audiological procedures could have been performed. These include threshold tone decay, auditory brainstem response (ABR) testing, and vestibular assessment. Most audiologists no longer perform the threshold tone decay or the rollover index tests because ABR tests and MRI scans have become more accessible.

Threshold tone decay testing is typically performed at a low frequency (e.g., 500 Hz) and a high frequency (e.g., 2000 Hz). Using the Olsen-Noffsinger procedure, the patient is asked to indicate how long he or she is able to sustain perception of the pure-tone stimuli presented for 60 seconds initially presented at 20 dB SL above the pure-tone threshold. If the patient is unable to perceive the stimulus for a full 60 seconds at 20 dB SL, the intensity is increased to 25 dB SL, and the 60 seconds cycle begins once again. This continues until the patient is able to sustain the signal for 60 seconds or reports that the signal is too loud. "Fast decay," or having little or no change in the length of perception as the signal is increased from 20 to 30 dB SL, suggests the presence of a retrocochlear lesion. A cochlear lesion is suggested if the length of time increases, suggesting "slow decay." Fast decay is associated with neural fatigue, which is highly abnormal, whereas slow decay is associated with normal neural adaptation. Although the threshold tone decay procedure is less sensitive than other procedures, it can be administered without special equipment and requires just a few minutes.

ABR tests reportedly have 96% sensitivity for detecting ANs larger than 1.5 cm. ABR results are positive for retrocochlear lesions if waves III and/or V are delayed or absent or if there is an increased interpeak latency. The sensitivity of the conventional ABR test to detect ANs smaller than 1.5 cm decreases to 70% In the case of small ANs, the summed response from all frequencies represented by the click stimulus is so strong that the impact of an AN infringing on a small section of the nerve is minimal and cannot be detected. In cases of normal ABR results, a stacked ABR test can be performed for further investigation. The stacked ABR test is more sensitive to detect small ANs because a derived-band technique is used to separate the contribution from each frequency region of the auditory nerve. Even a small AN would result in a reduced summed amplitude, making stacked ABR tests very sensitive to the presence of ANs smaller than 1.5 cm.

Vestibular assessment includes vestibular-evoked myogenic potentials (VEMPs) and the ENG and VNG test battery. VEMPs assess the inferior vestibular nerve. Approximately 80% of responses from patients with ANs have absent or decreased VEMP amplitudes. The bithermal caloric stimulation subtest of VNG assesses the superior vestibular nerve. A unilateral weakness may be found in patients with ANs, but the specificity of findings is low because reduced unilateral vestibular response can be associated with many other otologic conditions.

7.5.2 Radiology

Fig. 7.3 and Fig. 7.4 report the axial and coronal MRI findings, respectively, for BP. As indicated, the MRI scan confirmed the presence of a 1.5 cm AN pressing against the left middle cerebellar peduncle.

7.6 Diagnosis and Recommended Treatment

Treatment options for an AN include active surveillance, gamma knife radiation therapy, proton beam radiation, and microsurgical resection. Because ANs are typically benign and grow at a slow rate, active surveillance is a possible treatment plan. This approach involves routine serial radiographic imaging and audiograms. The schedule is typically every 6 months. Active surveillance is recommended by many physicians for ANs smaller than 2.5 mm or for adults over 65 years of age. Gamma knife radiation therapy is used to help prevent the AN from growing. Proton beam radiation has been recently introduced, and outcome reports have been quite favorable with respect to

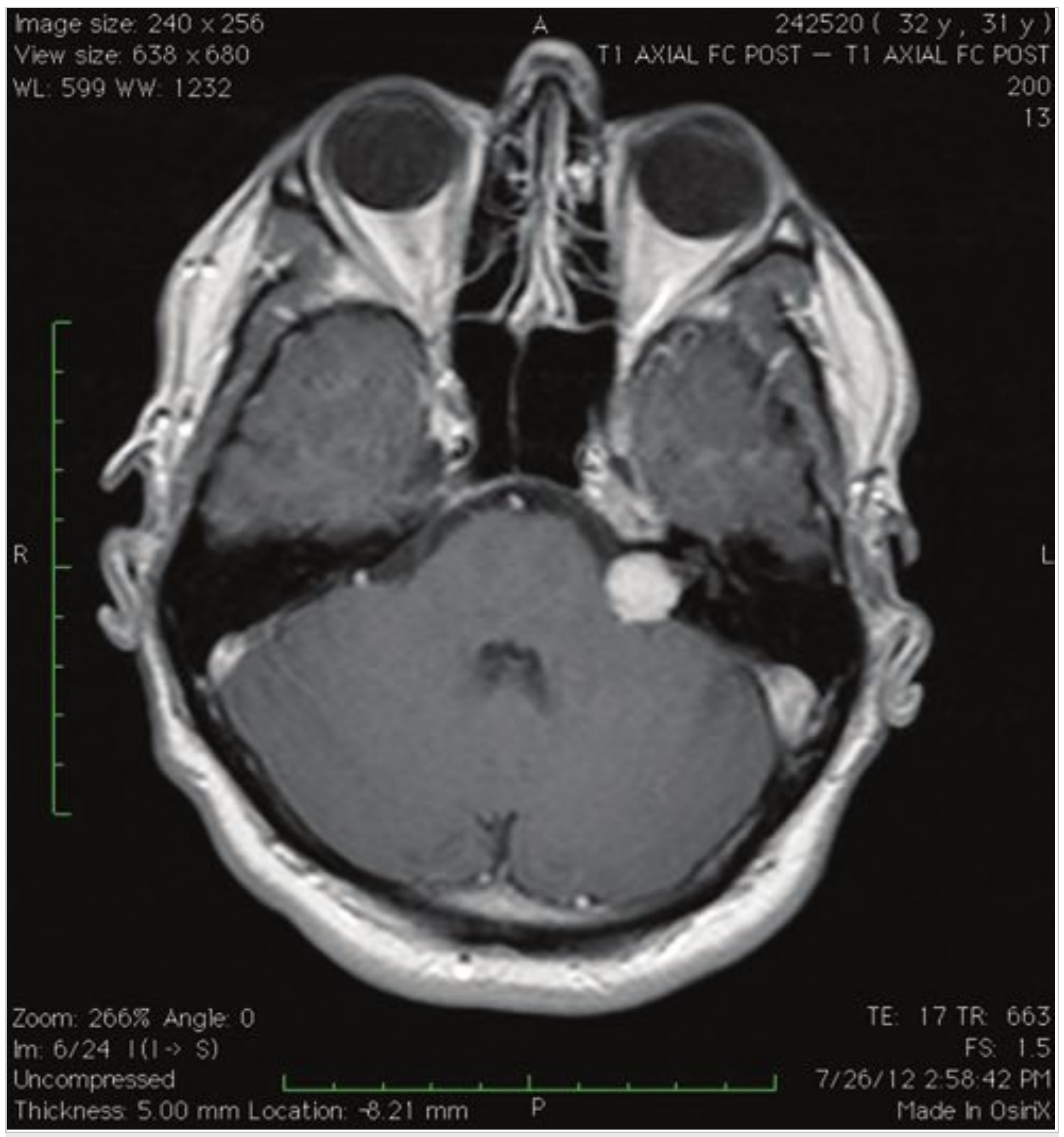


Fig. 7.3 Axial magnetic resonance imaging for BP.

preservation of pretreatment audiological and vestibular function with minimal side effects. Radiological treatment does not remove the AN but may reduce the size of the mass by interrupting the AN's deoxyribonucleic acid (DNA) reproduction. This approach is often recommended for patients with ANs smaller than 3.0 mm because of the potential for radiation damage to the surrounding structures.

Microsurgical resection can be used with all ANs but is more appropriate for patients whose ANs are large (>3 mm), have severe/profound hearing loss in the affected ear, or are experiencing neurological symptoms (e.g., paralysis, weakness, or spasms of facial nerve, headaches, changes in vision, or altered mental state) and dizziness. The surgical approach is either translabyrinthine, which involves drilling through the cochlear structures to access the AN mass, or via the posterior fossa approach that is less invasive to the inner ear structures and can preserve hearing in some cases. The surgeon makes the decision regarding the approach based on multiple factors. Important audiological factors for determining the surgical approach with possible hearing preservation include a pure-tone average below 50 dB HL, WRSs greater than 50%, normal ABR, and normal VEMP.

Risks of surgery include permanent or temporary facial nerve paralysis or weakness, balance or gait difficulties, and total hearing loss in the affected ear. If hearing cannot be preserved, surgical resection may cause a severe to profound unilateral sensorineural hearing loss, which is typically treated with bone conduction aids or bone-anchored hearing aids. These devices transmit sounds to the normal cochlea by vibrating the bones of the skull. The contralateral routing of the signal (CROS) amplification system is an alternative in which sound is transmitted from a microphone and processor worn on the poor ear to a receiver and open earmold on the normal ear.

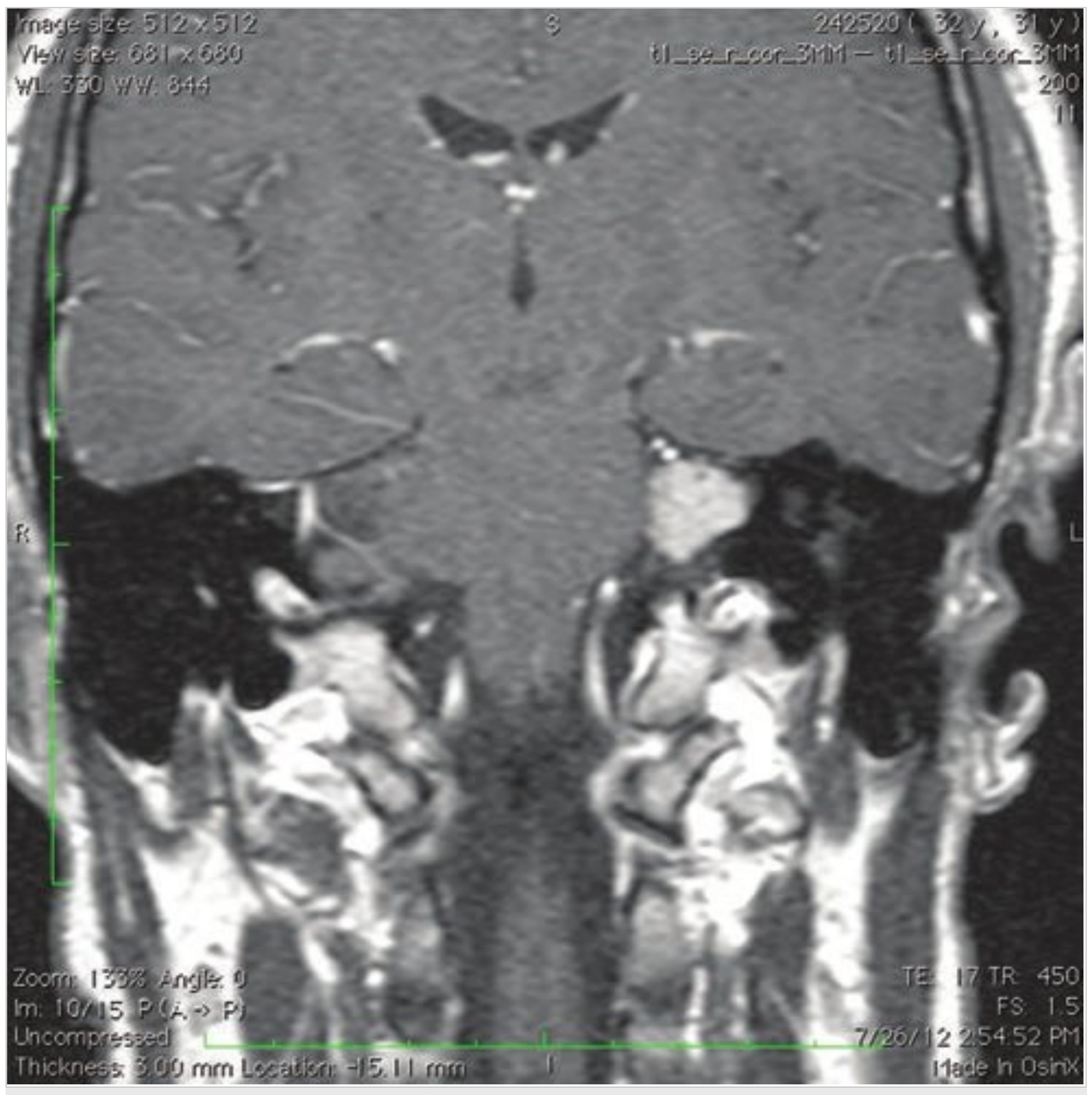


Fig. 7.4 Coronal magnetic resonance imaging for BP.

All of these options were presented to the patient who is currently undecided on which treatment option to pursue.

7.7 Key Points

1. Advanced audiological diagnostic procedures should be considered for all patients presenting with possible signs of AN. These include sudden or gradual unilateral hearing loss, tinnitus, WRS that is poorer than expected from the pure-tone audiogram, dizziness, or facial numbness or weakness.
2. Although an AN will be diagnosed by an MRI scan, audiological assessment is important for screening and determining treatment options and follow-up care.

7.8 Acknowledgments

The authors would like to thank the Dallas Ear Institute and Yoav Hahn, M.D., for the use of their MRI images, cooperation, and assistance.

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8 Clinical Management of a Patient with Bilateral Vestibular Schwannomas

Sarah O. Holbert, Michael J. Cevette, David Barrs, Linsey Scheibler, and Kristin Follett

A 75-year-old male with suspected neurofibromatosis type 2 (NF2) was clinically evaluated and subsequently decided to pursue cochlear implants (CIs).

8.1 Clinical History and Description

IJ is a 75-year-old male who presented to the otolaryngology department in May 2008 due to a sudden decrease in hearing in his left ear. IJ had also experienced sudden hearing loss in his right ear approximately 35 years prior. At that time, IJ was evaluated and was told he had a right vestibular schwannoma (VS), although he did not recall the details of the examination. IJ refused surgical removal of the VS and pursued no further subsequent testing. Since that time, the hearing in his left ear had been gradually decreasing until February 2008, when he suddenly lost all remaining hearing in his left ear. Since the decrease in hearing in the left ear in 2008, IJ had not been medically evaluated but had briefly tried bilateral hearing aids through a hearing aid dispenser, with no perceived benefit. IJ reported no history of dizziness or vertigo and denied otalgia, aural fullness, and any previous ear infections or surgeries. The otologist evaluated IJ and noted normal otoscopic findings. He was referred for audiological examination and magnetic resonance imaging (MRI) scan with gadolinium contrast to evaluate the presence of any retrocochlear lesion(s).

At the time of his audiological evaluation, IJ reported he occasionally had difficulty with imbalance, but confirmed that he had no prolonged dizziness or vertigo. He had some history of noise exposure including machinery and guns but had worn ear protection most of the time when shooting and when he was around machinery. Prior to his recent hearing aid trial, he had never used a hearing aid in the right ear due to limited benefit in functional speech recognition and had not used amplification in the left ear since 1989. Due to the recent decrease in hearing in the left ear, his only avenue to communicate with others had become written communication. Also, IJ had no ability to modulate the volume of his voice and was therefore shouting throughout the appointment.

8.2 Audiological Testing

The results of IJ's comprehensive audiological evaluation revealed bilateral profound sensorineural hearing loss (SNHL). IJ was unable to respond to any pure-tone signals at the limits of the audiometer. IJ's speech awareness threshold (SAT) was 100 dB HL in the left ear with no response in the right ear. IJ was provided information and brochures about CIs, and he expressed interest in pursuing this treatment option. Further testing and discussion of treatment options were delayed pending the results of his MRI scan.

8.3 Questions to the Reader

1. The otologist ordered an MRI scan with contrast to evaluate possible retrocochlear lesions. Which aspects of his case history and audiological examination could be consistent with a retrocochlear lesion?
2. Explain the expected impact of limited hearing aid use on the benefit from a CI.

8.4 Discussion of Questions to the Reader

1. The otologist ordered an MRI scan with contrast to evaluate possible retrocochlear lesion. Which aspects of his case history and audiological examination could be consistent with a retrocochlear lesion?
   IJ's report of a recent sudden hearing loss in his left ear, as well as a history of sudden loss and identification of a VS in his right ear, is consistent with a possible retrocochlear lesion. Other symptoms of a retrocochlear lesion may also include disequilibrium, tinnitus, trigeminal dysfunction, and facial neuropathy.
2. Explain the expected effects of limited hearing aid use on the benefit from a CI.
   Limited hearing aid use is believed to correlate ($p = \leq 0.05$) with poorer outcomes in speech recognition following the implantation of a CI. Other audiological factors, such as duration of the severe to profound hearing loss, are also correlated ($p = \leq 0.001$) with poorer outcomes with speech recognition with CIs.

8.5 Additional Testing

The MRI scan revealed a 1.8 cm mass that filled the left internal auditory canal and extended into the cerebellopontine angle. Results for the right side revealed a 4.4 mm intracanalicular enhancing mass with extension into the labyrinth (▶ Fig. 8.1). Both masses were consistent with VS. Due to the bilateral VS, it was suspected that the patient had neurofibromatosis type 2 (NF2). IJ's physical examination did not reveal any other hallmark signs such as subcutaneous nodules or neurofibromas, although several subcutaneous lipomas were identified across his body. IJ declined genetic testing to verify the NF2 diagnosis.

8.6 Diagnosis and Recommended Treatment

The patient met with the skull base tumor team (neurotology, neurosurgery, and radiation oncology) to review his options regarding the treatment of his tumors. The team recommended stereotactic radiosurgery or surgical removal, but IJ was not

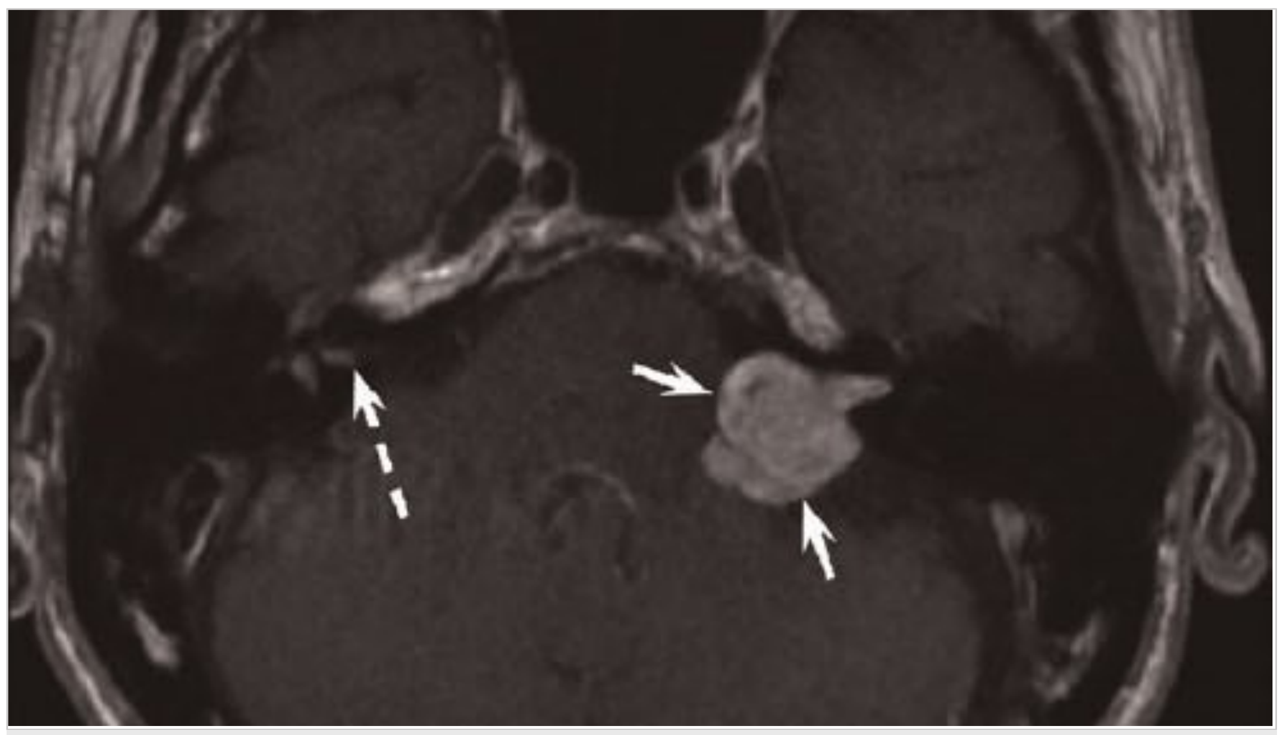

Fig. 8.1 Magnetic resonance imaging scan. axial, T1 with gadolinium contrast through the internal auditory canals showing the large enhancing left vestibular schwannoma (solid arrows) and the intracanalicular right VS (dashed arrow).

interested since it would not improve his hearing. Given his preferences and advanced age, an observational approach was undertaken to monitor tumor growth.

The otologist suggested he consider a CI for the right ear due to the small size of the VS and the appearance of an intact cochlear nerve. The left ear was not considered viable for a CI due to the size of the VS. The primary goal of this approach was to improve his hearing toward better communication with others and to improve his ability to modulate his loud vocal effort. An auditory brainstem implant (ABI) was not considered because tumor removal was not part of the treatment plan. The patient was cautioned regarding the potential for less than optimal speech recognition performance due to the existence of the tumor, the duration of the profound hearing loss in his right ear, and the lack of previous significant benefit from amplification. IJ was also cautioned that if the tumor on his right sidle enlarged after implantation, he could experience decreased benefit from the CI or even have to undergo explantation. After considerable counseling, IJ decided to pursue a right CI and was scheduled for a full CI candidacy evaluation to include aided speech testing, vestibular evaluation, and psychological assessment.

Aided speech recognition testing was completed with bilateral hearing aids programmed to National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) targets. Results revealed 0% aided sentence recognition for hearing in noise test (HINT) sentences presented in quiet at 70 dBA. His vestibular assessment included cervical vestibular-evoked myogenic potentials (cVEMPs) and videonystagmography (VNG). cVEMP responses were absent bilaterally when examined using a 500 Hz tone burst stimulus presented at 95 dB nHL. VNG results revealed no measurable caloric responses when tested with warm and ice water. This finding is suggestive of bilateral vestibular hypofunction. The remainder of the VNG battery was normal, including the oculomotor portion of testing, Dix-Hallpike maneuver, and static positional evaluation.

The patient's consultation with the cochlear implant team psychologist revealed minimal risk of depression, realistic expectations regarding potential benefit from a CI, and appropriate motivation to follow medical recommendations for follow-up. The psychologist also felt that IJ was prepared to deal with the disappointment if the surgery was not successful and that IJ acknowledged understanding about the risks of surgery.

Based on his candidacy evaluation, the CI team decided that IJ was a candidate for a CI in only the right ear.

8.7 Additional Questions to the Reader

1. Describe the typical clinical presentation and treatment options for NF2.
2. Separate Food and Drug Administration (FDA) criteria exist for eligibility for ABI and CI candidacy. Provide the specific distinctions between the FDA's ABI and CI candidacy as well as the expected outcome with each device.
3. This patient did not complete computerized dynamic posturography (CDP) during his candidacy evaluation; given his reported vestibular assessment results, however, what type of pattern could be expected on the sensory organization test (SOT)?

8.8 Discussion of Additional Questions to the Reader

1. Describe the typical clinical presentation and treatment options for NF2.
   NF2 is an autosomal dominant genetic disorder most often associated with bilateral VS. Symptoms may include gradual or sudden hearing loss, tinnitus, imbalance, as well as benign central and peripheral nervous system tumors. The onset of NF2 is typically during early adulthood. Regularly scheduled radiological monitoring is needed to assess tumor growth, and treatment may include observation, stereotactic radiotherapy, or surgical interventions, or a combination of all three depending on the size of the respective tumors. Management of hearing loss as a result of tumor growth and/or treatments includes hearing aids for those with aidable hearing and ABI for those with profound hearing loss. Although not the typical recommendation, use of a CI is also a treatment option that has recently been reported in the literature with results revealing a favorable outcome in speech recognition.
2. Separate Food and Drug Administration (FDA) criteria exist for eligibility for ABI and CI candidacy. Provide the specific distinctions between the FDA's ABI and CI candidacy as well as the expected outcome with each device (□ Table 8.1).
3. This patient did not complete computerized dynamic posturography (CDP) during his candidacy evaluation; given his reported vestibular assessment results, however, what type of pattern could be expected on the sensory organization test (SOT)?
   The SOT is part of the standard CDP test battery. During the SOT, patients are challenged to maintain balance under different conditions, such as when the visual surround and platform may be either fixed or sway-referenced. The movement of the platform and/or visual surround correspond to anterior-posterior sway movements, and the patient's eyes may be open or closed. With manipulation of these variables, the

Table 8.1 Description of Food and Drug Administration criteria and expected speech recognition of auditory brainstem implant (ABI) and cochlear implant (CI) recipients

FDA criteria and potential outcome	ABI	CI
FDA criteria	≥ 12 years of age - Postlingual HL - Diagnosis of NF2 with implantation occurs during tumor removal surgery - Approved in 2000 (FDA, 2013)	- Adults (18 +) with moderate to profound SNHL - Minimal benefit from appropriately fit hearing aids - 50% or less aided word recognition in the ear to be implanted and 60% in best aided condition - Children (1–17) with severe to profound SNHLLimited benefit with a trial with amplification - Approved in 1985 for adults and in 1990 for children ≥ 1 year of age
Potential outcome	- Pitch perception - Environmental awareness to sound - Aided speech reading - Limited open-set speech perception	- Pitch perception - Environmental awareness to sound - Aided speechreading - Limited to excellent open-set speech perception[a]

Abbreviations: NF2, neurofibromatosis type 2; SNHL, sensorineural hearing loss.
[a](Carlson et al, 2012).

SOT quantifies the roles of visual, vestibular, and somatosensory inputs in maintaining balance. Because IJ's VNG results suggested bilateral vestibular dysfunction, IJ would likely have difficulty maintaining his balance when both his vision and somatosensory inputs were disrupted. This would occur in Condition 5 (eyes closed, sway-referenced platform) and Condition 6 (sway-referenced visual surround and sway-referenced platform) of the SOT. Therefore, his SOT results would most likely have revealed a “5/6” pattern. This suggests that IJ would have scored below normal or would have fallen on Conditions 5 and 6. He would have been able to maintain his balance anytime his eyes were open with a stable visual reference or anytime he had a stable surface upon which to stand, as would be the case in Conditions 1 through 4. This information could be helpful in counseling the patient who may be at a greater risk of falling due to his vestibular dysfunction.

8.9 Outcome

A full electrode insertion of the right ear was achieved using a Cochlear Freedom Contour electrode array (Cochlea, Sydney, Australia). Placement was confirmed using a plain film X-ray. Intraoperative monitoring revealed normal impedance. Auto and advanced neural response telemetry (NRT) revealed no measurable response. NRT is an objective measure of the electrically evoked compound action potential (ECAP) from the auditory nerve, which can provide clinicians with information about an auditory system's response to electrical stimulation.

Comparisons of pre- and post-CI performance for aided sound-field frequency-modulated (FM) warble tone thresholds, HINT, and AzBio sentences revealed improved aided sound-field thresholds and significant improvement in speech recognition with the CI. Sentences presented alone in quiet, sentences mixed with 10-talker babble, and the warble tones were presented at 0° azimuth at 1 meter from IJ at ear level. The presentation level for the HINT and AzBio sentences was 60 dBA with a + 10 dB signal to noise ratio (SNR).

IJ's aided sound-field thresholds improved from “No Response” for unaided thresholds at the limits of the audiometer to aided thresholds of 20 to 25 dB HL with the CI. IJ's HINT score improved from 0% unaided to a ceiling aided performance of 88% within several months of activation. IJ's follow-up examination at his 5-year postactivation appointment revealed speech recognition performance at 81% on AzBio sentences in quiet and 37% on AzBio sentences mixed with 10-talker babble presented at a + 10 SNR (Fig. 8.2). A follow-up computed tomographic (CT) scan with contrast at 5 years after the diagno-

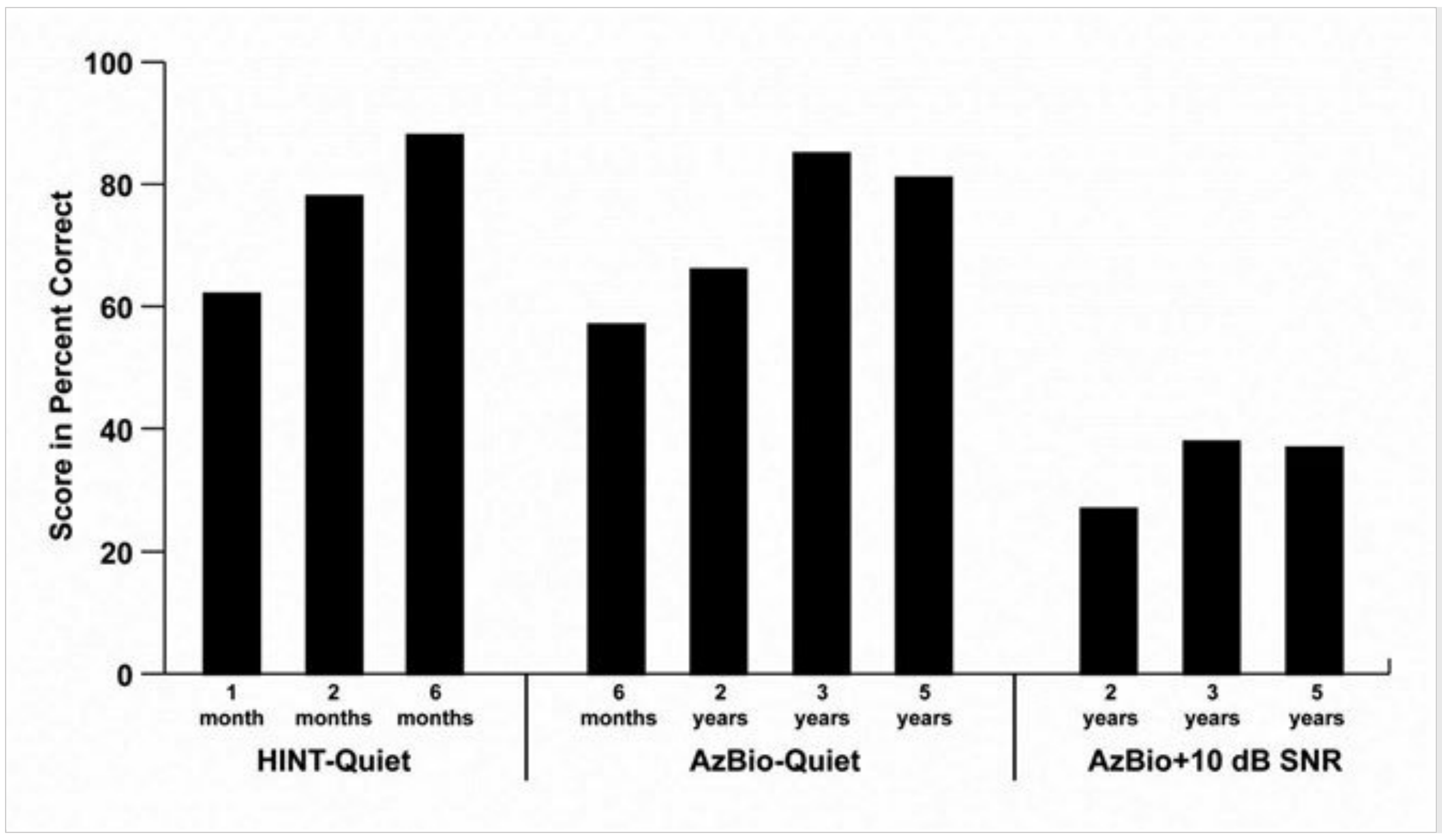

Fig. 8.2 Speech recognition testing at 5 years following the implantation of the cochlear implant. The patient's 5-year postactivation speech recognition performance on AzBio sentences in quiet was 81% and 37% at a + 10 dB signal to noise ratio.

sis did not demonstrate any tumor growth. In some instances, an MRI scan may be ordered to further define the extent of tumor growth. MRI is rarely used when a CI is present because the CI magnet must be surgically removed in order for this to take place. The removal of the CI magnet would involve the risks associated with administering general anesthesia and possible damage to the CI. Also, it is possible to complete an MRI scan for a patient with a CI, but it would require lowering the strength of the MRI magnet used in order to obtain a good image. This could lead to less than optimal images if the magnet is left in place.

IJ's listening program, or MAP, used Cochlear Corporation's default settings. A MAP is a set of parameters including rate, pulse width, and maxima that constitute a listening program. Clinicians create different MAPs in order to optimize patients' speech recognition abilities. Impedances have remained stable over 5 years, with the exception of two electrodes (16 and 19) that were identified as having a short circuit in 2013. The patient also had NRT responses with normal morphology after activation using AutoNRT or the default test parameters in the Cochlear Software.

CIs may be an option for some patients with NF2, even when radiological evidence indicates extensive, or long-standing, tumor involvement. Given IJ's advanced age (75 years); prior poor response to amplification; and size, location, and growth history of his tumors, the CI was determined to be the best course of treatment for IJ. This recommendation was felt to address his hearing needs and maximize his potential for improved communication. IJ was able to obtain significant open-set word recognition and improved voice modulation through the use of the CI. IJ is no longer using writing as the primary mode of communication and is able to effectively use the telephone.

8.10 Additional Questions to the Reader

1. NRT during surgery did not initially yield measurable responses. Is this predictive for IJ's outcome?
2. Both MRI and CT may be used with CI patients; however, different generations of internal devices have differing criteria for testing with MRI. Create a table listing compatibility for the internal devices of Advanced Bionics (Valencia, CA), Cochlear Corporation, and Med-El Corporation (Durham, NC).

8.11 Discussion of Additional Questions to the Reader

1. NRT during surgery did not initially yield measurable responses. Is this predictive for IJ's outcome?
   Limited research has focused upon outcomes of patients with no measurable ECAP thresholds. The presence, however, of these responses is generally believed to be a positive sign of nerve function and the outcome of improved speech recognition.

Table 8.2 Food and Drug Administration–approved magnetic resonance imaging (MRI) compatibility for cochlear implant (CI) internal device

Advanced Bionics internal devices	FDA-approved MRI compatibility
HiRes 90K Series	0.3 or 1.5 Tesla with magnet removed (Advanced Bionics 90K and 90K Advantage, 2013)
CII Series	Not Approved
C1 Series	Not Approved
Cochlear[b] internal devices	FDA-approved MRI compatibility (Cochlear Physicians Package Insert, 2010)
CI500 Series	1.5 Tesla or less with magnet removed
Freedom Series	1.5 Tesla or less with magnet removed
Nucleus 24 Series	1.5 Tesla or less with magnet removed
Nucleus 22 Series	1.5 Tesla or less for internal devices with magnet removed (those with C, G, H, J, L, P, T, 2, 5, or 7 as the middle character printed on the internal implant) Contraindicated for recipients without removable magnet and a middle letter on internal device other than those listed above
Med-EL[c] internal devices	FDA-approved MRI compatibility(Med El Press release 6/2013)
Concert Series	1.5 Tesla or less with magnet in place
Sonata Series	1.5 Tesla or less with magnet in place
Pulsar Series	1.5 Tesla or less with magnet in place
Combi 40 Series	1.5 Tesla or less with magnet in place

[a] Advanced Bionics, LLC, Valencia, CA.
[b] Cochlear, Centennial, CO.
[c] Med-El Corporation, USA, Durham, NC.

2. Both MRI and CT may be used with CI patients; however, different generations of internal devices have differing criteria for testing with MRI. Create a table listing compatibility for internal devices of Advanced Bionics (Valencia, CA), Cochlear Corporation, Med-El (Durham, NC).
   (Table 8.2)

8.12 Key Points

1. Careful consideration of clinical findings, case history, and patient needs is necessary for a successful outcome. Given IJ's stable tumor growth and advanced age, CI was an option that also provided the best opportunity to improve IJ's speech recognition despite the diagnosis of NF2.
2. Although the primary intent of CIs is to restore hearing due to cochlear hearing loss, this case report provides supporting evidence that even those patients who display VS may be successful CI recipients.

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9 An Unexpected Case of Otosclerosis

Kristi Oeding

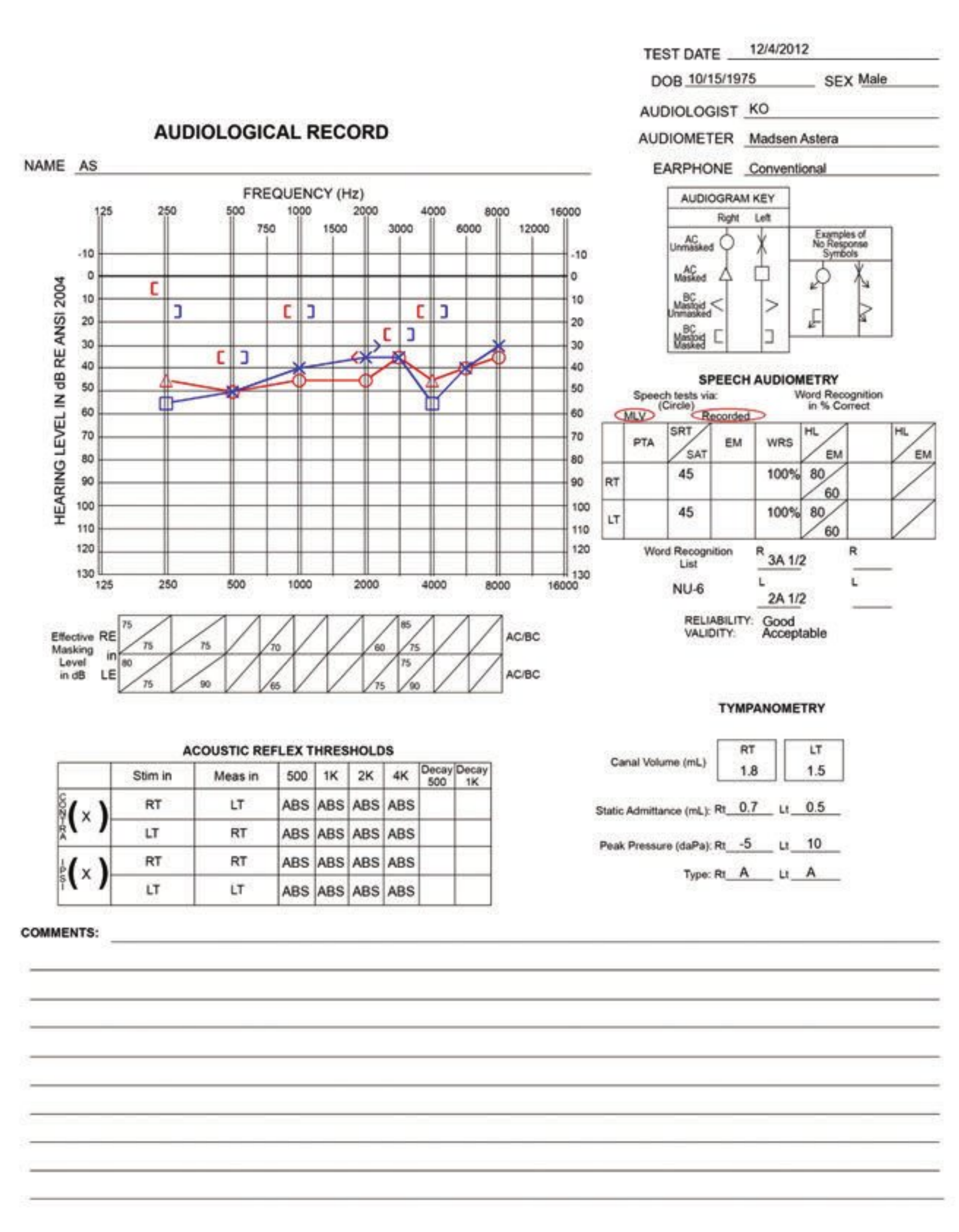
A patient presents with symptoms of gradual hearing loss over the past year and reports increased difficulty hearing after recent bilateral ear infections. Results of an audiological and otologic examination revealed classic signs of otosclerosis.

9.1 Clinical History and Description

AS, a 37-year-old African American male, reported decreased hearing bilaterally over the past year that he attributed to a virus resulting in bilateral otitis media. He reported that a prior audiological examination revealed that his hearing had decreased 20 dB HL below the range of normal hearing. AS reports that he hears better on some days than on other days and that his wife tells him he plays the television too loud. AS notices that he can "hear" speech, but cannot always "understand" what is said. He also reported constant bilateral tinnitus for the past 3 years, an occasional feeling of being "off-balance," a family history of hearing loss due to aging, and occupational noise exposure from working as a forklift operator in an automobile factory and the airport for more than 10 years with limited use of hearing protection.

9.2 Audiological Testing

Otoscopy was unremarkable. An audiological examination (Fig. 9.1) revealed bilateral symmetric moderate rising to a mild mixed hearing loss from 250 to 3000 Hz, sloping to a moderate conductive hearing loss at 4000 Hz, and rising to a mild hearing loss from 6000 to 8000 Hz. Speech recognition thresholds (SRTs) revealed a moderate loss in the ability to receive speech bilaterally. Word recognition scores (WRSs) using Northwestern University Auditory Test No. 6 (NU-6) word lists with a recorded female talker at the patient's most intelligible level revealed normal ability to recognize speech bilaterally.



AUDIOLOGICAL RECORD

TEST DATE 12/4/2012
DOB 10/15/1975 SEX Male
AUDIOLOGIST KO
AUDIOMETER Madsen Astera
EARPHONE Conventional
NAME AS

SPEECH AUDIOMETRY

Speech tests via: (Circle) [✓ MLV] [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT		45		100%	80/60		
LT		45		100%	80/60		

Word Recognition List NU-6
R 3A 1/2 R
L 2A 1/2 L

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB

RE in	75/75	/75		/70			/60	85/75			AC/BC
LE in	80/75	/90		/65			/75	75/90			AC/BC

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.8	1.5

Static Admittance (mL): Rt 0.7 Lt 0.5
Peak Pressure (daPa): Rt -5 Lt 10
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	ABS	ABS	ABS	ABS		
IPSI (X)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS:

Fig. 9.1 Patient's comprehensive audiological and immittance examination.

Immittance audiometry was performed and revealed normal tympanograms bilaterally and absent acoustic reflex thresholds (ARTs) at 500 to 4000 Hz to ipsilateral and contralateral stimulation bilaterally.

9.3 Questions to the Reader

1. Based on the case history, what etiology and/or audiology result would you expect?
2. Based on the comprehensive audiological examination in Fig. 9.1, what are some possible causes of this type and configuration of hearing loss?
3. What other evaluation tools could be used to further diagnose the patient as a result of the comprehensive audiological examination?
4. Based on the results of immittance audiometry, what could be the possible cause of hearing loss?
5. What are some common audiological examination tools and results that aid in the diagnosis of otosclerosis?
6. What patient population would typically be expected to be diagnosed with otosclerosis?
7. Based on these test results, what recommendations should be made?

9.4 Discussion of Questions to the Reader

1. Based on the case history, what etiology and/or audiological result would you expect?
   Several types of hearing loss could be probable from the case history, including a sensorineural component due to the patient's history of gradual noise exposure or due to his recent viral infection. Another possibility would be a conductive component due to the patient's recent report of bilateral otitis media.
2. Based on the comprehensive audiological examination in Fig. 9.1, what are some possible causes of this type and configuration of hearing loss?
   Possible factors that could be causing the hearing loss revealed as a result of the comprehensive audiological examination could include recurrent otitis media. The patient's report of a more than 10-year history of noise exposure with limited use of hearing protection could help explain the sensorineural component at 2000 and 3000 Hz. Finally, the presence of the Carhart notch for bone-conduction thresholds at 2000 Hz could indicate possible otosclerosis. The Carhart notch is typically a sensorineural hearing loss at 2000 Hz, while the surrounding frequencies have a conductive component. The proposed reason for the 2000 Hz notch is due to the loss of the resonance of the ossicular chain at 2000 Hz due to the fixation of the stapes footplate.
3. What other evaluation tools could be used to further diagnose the patient as a result of the comprehensive audiological examination?
   Immittance audiometry is warranted to further examine the conductive component of the patient's hearing loss.
4. Based on the results of immittance audiometry, what could be the possible cause of hearing loss?
   Otitis media is less likely an etiology of the patient's hearing loss given the normal tympanograms. On the other hand, otosclerosis is more likely due to the normal tympanograms and absent ARTs to ipsilateral and contralateral stimulation.
5. What are some common audiological examination tools and results that aid in the diagnosis of otosclerosis?
   The following are possible indicators of otosclerosis:
   a) Case history: Family history of otosclerosis (approximately 40% with the genotype develop otosclerosis), occasional vertigo (approximately 25% of patients), and tinnitus (approximately 75% of patients) can be indicators of otosclerosis.
   b) Otoscopy: The Schwartze sign may be seen as a reddish hue due to the vascularization of the spongy bone and is present in approximately 10% of patients with otosclerosis.
   c) Audiological examination: Typically, a conductive or mixed hearing loss is present in the low frequencies and can advance to the high frequencies as otosclerosis progresses, with the exception of a bone-conduction "Carhart notch" that is typically seen at 2000 Hz resulting in a sensorineural hearing loss. The Carhart notch has been reported to be present in about 73 to 80% of patients with otosclerosis.
   d) Immittance audiometry: Hypocompliant (<0.2 mL), normal (0.3–1.5 mL), or hypercompliant (>1.5 mL) tympanograms along with elevated or absent ARTs may be seen depending on the progression of otosclerosis. Approximately 62 to 72% of patients with otosclerosis can have normal tympanograms, approximately 20 to 38% hypocompliant tympanograms, and approximately 8% hypercompliant tympanograms. As can be seen, the results of tympanometry are variable.
6. What patient population would typically be expected to be diagnosed with otosclerosis?
   Otosclerosis is twice as likely to be seen in females than males and is bilateral in 70 to 80% of cases. Onset is between ages 15 and 45 years. Also, otosclerosis is more prevalent in Caucasians (approximately 0.04–1%). Although the cause of otosclerosis remains unclear, some theories are related to genetics (autosomal dominant inheritance), viruses (particularly measles), involvement of the endocrine and immune systems, bone remodeling, and connective tissue disorders. Also, several studies have shown that pregnancy may increase the progression of otosclerosis.
7. Based on these test results, what recommendations should be made?
   Due to the mixed hearing loss, referral to an otologist is recommended to determine possible medical remediation of the hearing loss. If the otologist determines the diagnosis is otosclerosis a stapedotomy or stapedectomy may be performed. A stapedotomy involves creating a small hole in the stapes footplate, whereas a stapedectomy involves partial or complete removal of the stapes footplate prior to the placement of the prosthesis. Audiological examination after medical management or annual audiological examinations are recommended. Amplification is also a treatment option after medical management is completed, medical clearance is obtained, and if the patient is motivated toward amplification.

9.5 Diagnosis and Recommended Treatment

AS was scheduled to be seen by an otologist the same day of the audiological examination. The otologist reported that microscopic otoscopy of the tympanic membranes revealed bilateral Schwartze signs. The otologist scheduled to see AS 1 month later after the patient had a computed tomographic (CT) scan, which revealed no abnormalities of the temporal bone, but the otologist noted questionable oval window otosclerosis bilaterally. Based on the results of otoscopy, the audiological examination, and the CT scan the otologist reported that AS had bilateral otosclerosis. After discussing the treatment options of surgery and/or amplification, the patient preferred to have surgery on the left ear because he perceived greater difficulty hearing from this ear.

9.6 Outcome

An appointment was scheduled for a left stapedotomy, and the surgery was performed without complications. AS experienced some initial mild imbalance postsurgery, but the imbalance subsided by the 1 month follow-up. AS noted such a marked improvement in hearing in the left ear that he did not want surgery on the right ear. Unfortunately, AS was not scheduled for an audiological examination on the day of his follow-up with his otologist, and he did not return for the 3-month follow-up appointment; therefore, the extent of improvement in his hearing is unknown.

9.7 Key Points

1. This case highlights the importance of performing audiological and immittance examinations in the differential diagnosis of hearing loss.
2. Some classic signs of otosclerosis are family history, Schwartze sign, Carhart notch, and hypocompliant or normal tympanograms with elevated or absent ARTs.
3. Although more common in women and Caucasians, otosclerosis, although rare, can also affect men and persons of African descent and is typically present bilaterally.
4. It is important to have an interdisciplinary team of health-care professionals in the management of patients to differentially diagnose hearing disorders.

Suggested Reading

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10 A Case of Mistaken Identity

Judy L. Peterein

This case report outlines the evaluation and treatment options for a patient previously diagnosed with Ménière disease. The patient was evaluated and differentially diagnosed based on history, clinical examination, audiological and acoustic immittance test results, electrocochleography (ECoG), vestibular evaluation, and spiral computed tomographic (CT) scans.

10.1 Clinical History and Description

SC was 65 years of age when she was first seen in the audiology clinic. She had been seen at another large medical center for evaluation 7 months prior and had been diagnosed with left Ménière disease. She reported a 20-year history of fullness and ringing in the left ear as well as imbalance. At the time of her initial visit to this clinic SC reported a feeling of constant imbalance and constant pressure in her left ear. She also reported hearing loss in her left ear. In addition, she reported that her eyes "jump" when she hears loud sound in her right ear such as the telephone ringing. Finally, she stated she feels a rattle or vibration in her head when she talks.

SC was seen by an otologist at this clinic who specializes in balance disorders. Physical examination in the office revealed increased sway on gait and tandem Romberg testing. A positive superior canal sign (upward and intorsional nystagmus) was noted on the right side with pressure applied to the right tympanic membrane via pneumatic otoscopy. Based on SC's history and physical examination, the otologist ordered several diagnostic tests including comprehensive audiometry, acoustic immittance, and ECoG. A battery of vestibular tests was also ordered including video-oculography (VOG), rotational chair testing, and computerized dynamic posturography (CDP) testing. A spiral CT scan was also ordered.

10.2 Audiological Testing

A comprehensive audiological examination revealed a mild to moderate mixed hearing loss through 1000 Hz rising to a slight loss from 2000 to 6000 Hz then rising to normal hearing at 8000 Hz in the right ear. Significant air–bone gaps (15–20 dB HL) were noted at 250, 500, and 1000 Hz. Pure-tone test results for the left ear revealed a slight to mild mixed hearing loss through 4000 Hz sloping to a moderate loss at 6000 Hz and a moderately severe loss at 8000 Hz. A 30 dB air–bone gap was noted at 250 Hz. Speech recognition thresholds (SRTs) were mildly impaired bilaterally (35 and 30 dB HL). Word recognition scores (WRSs) were within normal limits bilaterally (96% and 100%). Acoustic immittance testing revealed tympanograms to be within normal limits bilaterally, suggesting normal middle ear function. Right and left ipsilateral acoustic reflex thresholds (ARTs) were normal. Right contralateral ARTs were normal at 1000, 2000, and 4000 Hz and slightly elevated at 500 Hz. Left contralateral ARTs were normal at 500, 1000, and 2000 Hz and slightly elevated at 4000 Hz. Acoustic reflex decay was normal at 500 and 1000 Hz stimulating the left ear and normal at 1000 Hz stimulating the right ear. Reflex decay could not be tested at 500 Hz in the right ear due to the elevated ART at 500 Hz (□ Fig. 10.1).

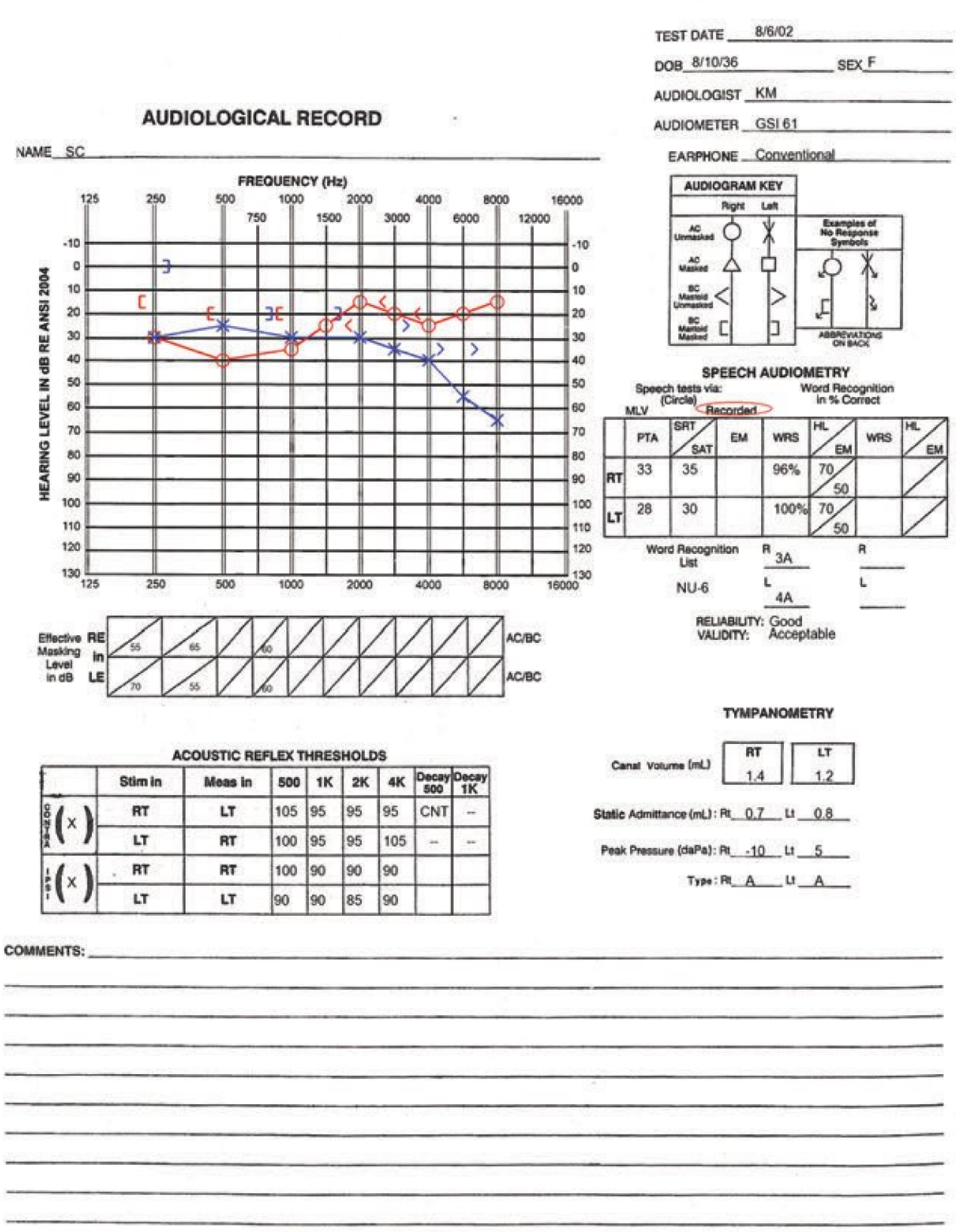
ECoG testing was performed using a gold wrapped tiptrode electrode as the primary recording electrode with reference electrodes placed at both the vertex and the earlobe contralateral to stimulation. Alternating polarity acoustic click stimuli were delivered via Nicolet Bionedical Model TIP 300 insert receivers at stimulus presentation levels of 85 and 95 dB nHL. A recording time window of 10 milliseconds was utilized with a band pass filter setting of 5 to 3000 Hz. Summating potential (SP)/action potential (AP) amplitude ratios were calculated for both sides and were enlarged (>0.5), bilaterally. Average SP/AP amplitude ratios of 0.71 and 0.76 were obtained for the left and right sides, respectively (□ Fig. 10.2 and □ Fig. 10.3).

10.3 Questions to the Reader

1. Based on the patient's case history is a diagnosis of Ménière disease suggested?
2. How is it possible to have the presence of significant air–bone gaps and present acoustic reflexes?
3. What do the enlarged SP/AP amplitude ratios obtained via ECoG suggest?

10.4 Discussion of Questions to the Reader

1. Based on the patient's case history is a diagnosis of Ménière disease suggested?
   SC does not demonstrate the classic symptoms of Ménière disease/syndrome. Ménière disease is characterized by four symptoms, including fluctuating hearing loss, tinnitus, aural fullness, and vertigo. In addition, symptoms occur in spells or attacks often beginning with unilateral aural fullness and low pitch roaring tinnitus then developing into hearing loss in the symptomatic ear followed by vertigo lasting anywhere from 30 minutes to 2 hours. After the vertigo spell resolves, hearing loss, tinnitus, and aural fullness also resolve. A general feeling of disequilibrium, however, may remain for a few days. Eventually, all symptoms will resolve and the patient will have no symptoms until the next episode. Although SC did report left tinnitus, aural fullness, and hearing loss, the symptoms were fairly constant. She did not report fluctuation in hearing. She also did not describe the spells of vertigo associated with Ménière disease, but rather a constant feeling of imbalance.
2. How is it possible to have the presence of significant air–bone gaps and present acoustic reflexes?
   In general, when significant air–bone gaps are present on the audiogram, ARTs are expected to be absent at least when measuring the reflex in the ear with the air–bone gap. In SC's left ear, a 30 dB air–bone gap is noted at 250 Hz. Air–bone



TEST DATE 8/6/02
DOB 8/10/36 SEX F
AUDIOLOGIST KM
AUDIOMETER GSI 61
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME SC

FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

Effective Masking Level in dB											
RE in	55	65		60							AC/BC
LE	70	55		60							AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	33	35		96%	70 / 50		
LT	28	30		100%	70 / 50		

Word Recognition List R 3A R
NU-6 L 4A L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.4	1.2

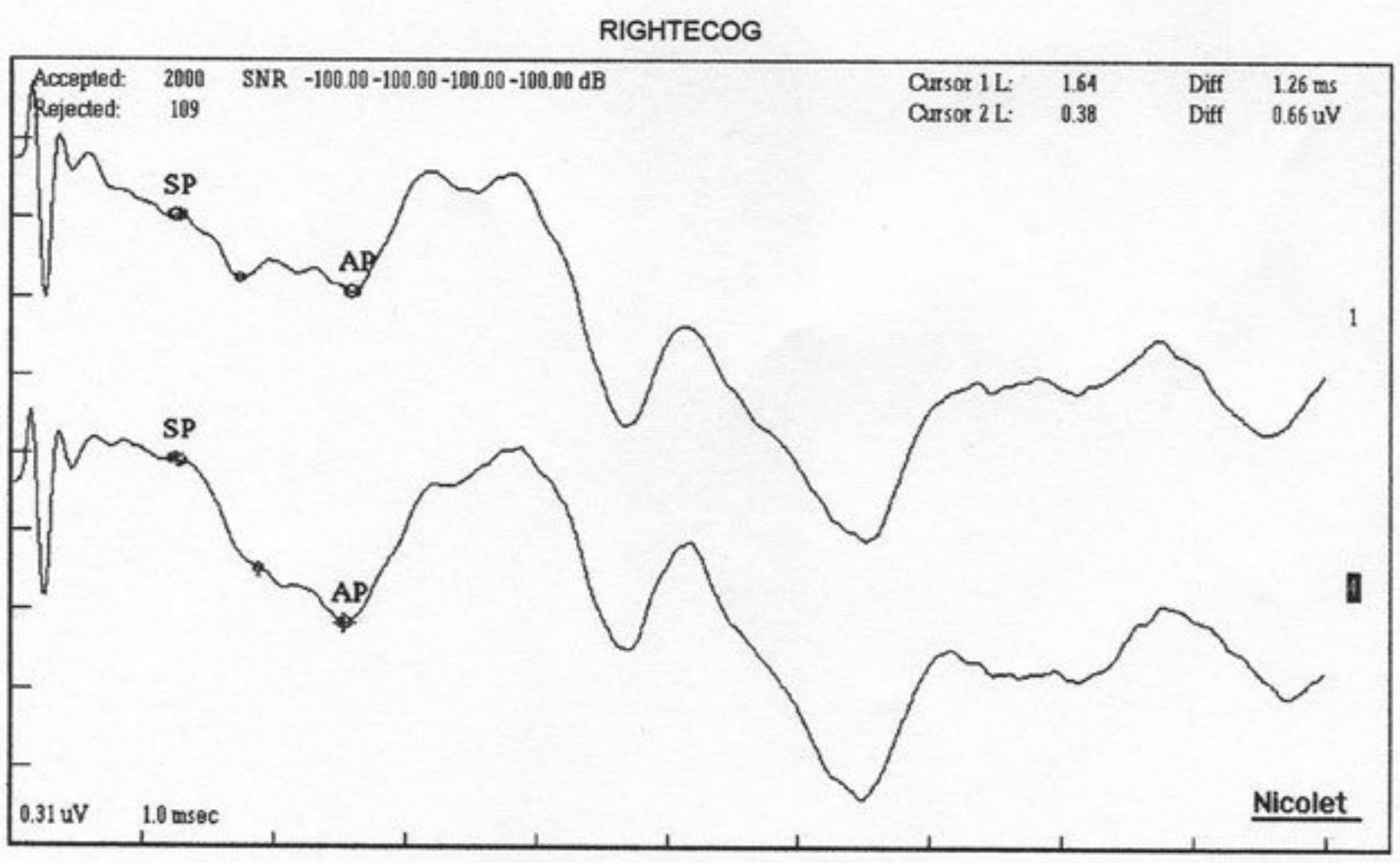
Static Admittance (mL): Rt 0.7 Lt 0.8
Peak Pressure (daPa): Rt -10 Lt 5
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	105	95	95	95	CNT	--
	LT	RT	100	95	95	105	--	--
IPSI (X)	RT	RT	100	90	90	90		
	LT	LT	90	90	85	90		

COMMENTS:

Fig. 10.1 Comprehensive audiological examination completed at initial appointment.



RIGHTECOG

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Accepted:  2000   SNR  -100.00 -100.00 -100.00 -100.00 dB      Cursor 1 L:  1.64    Diff  1.26 ms
Rejected:  109                                                 Cursor 2 L:  0.38    Diff  0.66 uV
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Fig. 10.2 Right electrocochleography waveforms—average summating potential (SP)/action potential (AP) amplitude ratios = 0.76.

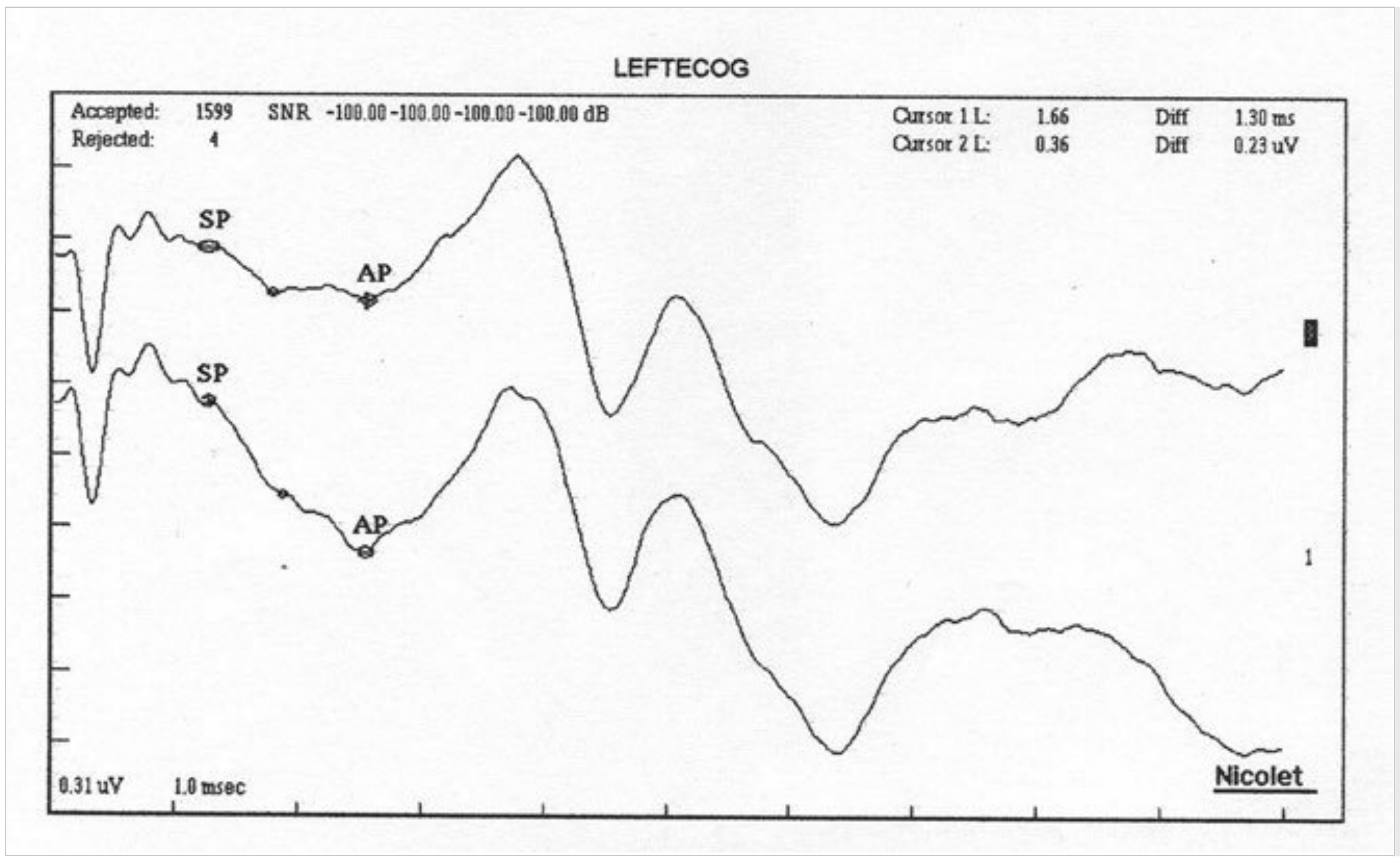

Fig. 10.3 Left electrocochleography waveforms—average summating potential (SP)/action potential (AP) amplitude ratios = 0.71.

gaps (15–20 dB HL) were noted at 250, 500, and 1000 Hz in the right ear. ARTs are present in all test conditions. One explanation for the presence of the ARTs is the possibility of a "third mobile window" created by a dehiscence in one of the semicircular canals. Because the dehiscence accentuates the difference in compliance between the scala tympani and scala vestibuli, the sensitivity for bone conducted sound may be better than normal or better than expected. In cases of superior semicircular canal dehiscence (SSCD), the middle ear may be entirely normal, but due to better than expected bone conduction sensitivity, air–bone gaps are present. The 0 dB HL bone conduction threshold at 250 Hz in the left ear supports this point. Acoustic immittance testing may initially suggest ossicular fixation until ART is completed and ARTs are present. The presence of normal tympanograms, present ARTs, and significant air–bone gaps are consistent with the third mobile window created by the bony dehiscence. Better than expected hearing via bone conduction may also provide an explanation for the rattle or vibration SC feels in her head while talking.

3. What do the enlarged SP/AP amplitude ratios obtained via ECoG suggest?
   An enlarged SP/AP amplitude ratio can suggest more than one underlying inner ear etiology. The most extensively researched pathological processes involving the inner ear with associated enlargement of the SP/AP amplitude ratio are Ménière disease and endolymphatic hydrops. Many investigators have established a positive correlation between an enlarged SP/AP amplitude ratio and Ménière disease and endolymphatic hydrops. An enlarged SP/AP amplitude ratio can also be recorded in some cases of perilymphatic fistula. In the current case report, however, the most likely underlying cause for bilaterally enlarged SP/AP amplitude ratios is the presence of SSCD, bilaterally. Several studies support the presence of enlarged SP/AP amplitude ratios in cases of SSCD. In cases of Ménière disease and hydrops as well as perilymphatic fistula, unilateral findings would most often be expected, although bilateral Ménière disease is a possibility. SC's history, clinical examination, audiological findings, and ECoG test results support the diagnosis of bilateral SSCD.

   Vestibular test results and spiral CT findings will be discussed in the following section.

10.5 Additional Testing

Results for the battery of vestibular tests were as follows. Standard VOG test results were within normal limits. Bithermal caloric results were symmetric with only an 8% left relative reduced vestibular response with a total eye speed of 83 deg/s. Rotational chair testing was within normal limits with normal vestibular ocular reflexes (VORs), normal step velocity time constants per, rotation and post rotation and normal fixation suppression. CDP revealed decreased vestibular utilization. Decreased scores on sensory organization test (SOT) conditions 5 and 6 with late falls is consistent with a vestibular dysfunction pattern and is seen with either reduced bilateral peripheral vestibular function, uncompensated unilateral loss of peripheral vestibular function, or poor utilization of vestibular cues. Motor control, strategy analysis, center of gravity alignment, and adaptation were all within normal limits.

Spiral CT scan, in the orthogonal superior semicircular canal plane, showed dehiscence of the superior canal, bilaterally.

10.6 Additional Questions to the Reader

1. Did the battery of vestibular tests contribute significantly to the diagnosis of SSCD?
2. What other test(s) might have provided useful diagnostic information regarding the presence of SSCD?

10.7 Discussion of Additional Questions to the Reader

1. Did the battery of vestibular tests contribute significantly to the diagnosis of SSCD?
   Overall, findings on vestibular testing were normal. This is not unusual in cases of semicircular canal dehiscence. VOG

recordings, however, during delivery of loud sound or pressure to the affected ear may result in a characteristic upward and intorsional nystagmus associated with SSCD. This finding was noted during the clinical examination by the otologist. When pressure was applied to the right ear via pneumatic otoscopy, elevation and intorsion of the eyes were observed.

2. What other test(s) might have provided useful diagnostic information regarding the presence of SSCD?
   Vestibular-evoked myogenic potential (VEMP) testing is an important part of the vestibular test battery in the diagnosis of SSCD. Most peripheral diseases affecting the vestibular system result in decreased VEMP amplitudes or absence of the VEMP. In contrast to these findings, enlarged VEMPs with lower VEMP thresholds are found in the case of SSCD. Hypersensitivity to sound stimulation in SSCD reflected in the described VEMP findings is due to the third window created by the exposed membrane of the superior semicircular canal. Whereas sound normally enters the inner ear via the oval window with dissipation of sound pressure by movement of the round window, in cases of SSCD the third window allows sound to be dissipated through the dehiscence and thus bypass the cochlea. As part of our standard test protocol VEMP testing is routinely performed in our clinic in cases of suspected semicircular canal dehiscence.

10.8 Diagnosis and Recommended Treatment

After reviewing the patient's case history and the extensive battery of diagnostic tests completed, the final diagnosis for SC was bilateral SSCD. The patient did not have Ménière disease as was previously diagnosed at another facility. Medical/surgical treatment options recommended by the otologist were myringotomy and tube placement for possible sound damping and/or possible sound cancellation, sound-attenuating ear plugs, or a middle fossa approach for superior canal plugging. In addition, hearing aids were recommended due to bilateral hearing loss. SC returned to the clinic to discuss hearing aid options. She was not enthusiastic about the option of hearing aids. In fact, she was concerned about the use of hearing aids because her eyes move in response to loud sound. She also did not feel that she had significant difficulty hearing. The only situations in which she noticed difficulty hearing were with her soft spoken brother and at times in group listening. Family members who attended the hearing aid appointment encouraged her to try hearing aids. SC decided that she would try a hearing aid only in her left ear and was subsequently fit with a 15-channel digital behind-the-ear (BTE) hearing aid coupled to the left ear with a slim tube and an open dome. Hearing aid performance was verified using real ear measures. A Frye 6500 real ear analyzer (Frye Electronics, Inc., Tigard, OR) was used to verify that the hearing aid settings provided gain that met the National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) prescriptive target for an input level of 65 dB sound pressure level (SPL). In addition, input levels of 50 and 80 dB SPL were also measured to verify the nonlinear performance of the hearing aid. Directional microphone performance was then verified using real ear measures. SC purchased the hearing aid on a 30-day trial.

10.9 Outcome

Extensive diagnostic testing confirmed the presence of bilateral SSCD. Although several treatment options were offered to SC, she elected not to undergo medical or surgical treatment. Due to the presence of bilateral mixed hearing loss, hearing aids were also offered as an option to improve her hearing. SC purchased a hearing aid for her left ear due to strong encouragement from her family. She did not wear the hearing aid, and it was returned in the mail approximately 2 weeks after the purchase date.

Although SC did not elect to have medical or surgical treatment at the time of diagnosis of bilateral SSCD, she may wish to do so in the future. It was recommended that she return for follow-up with the otologist in 1 year and as needed. Audiological follow-up was recommended in 1 year. It is possible that SC may be interested in hearing aids in the future.

10.10 Key Points

1. Careful evaluation of case history information is important in the diagnosis of patients with balance disorders.
2. Previous diagnoses must be carefully considered with appropriate follow-up evaluation when patients are seen in the clinic for the first time.
3. Symptoms that may indicate the presence of SSCD include the following:
   a) Eye movement in response to either loud sound or increased intra- or intercranial pressure
   b) Dizziness or vertigo in response to either loud sound or increased intra- or intercranial pressure
   c) Autophony
4. Diagnostic test results suggesting the presence of SSCD include the following:
   a) Better than expected bone conduction thresholds
   b) Low-frequency air–bone gaps on the audiogram with normal tympanometry and present ARTs
   c) Enlarged SP/AP amplitude ratios on ECoG
   d) VOG recordings during delivery of loud sound or pressure to the affected ear resulting in an upward and intorsional nystagmus
   e) Large-amplitude VEMPs with lower than normal thresholds
   f) Visualization of the areas of bony dehiscence by spiral CT scans

Suggested Reading

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11 Gamma Knife Treatment for Acoustic Tumor

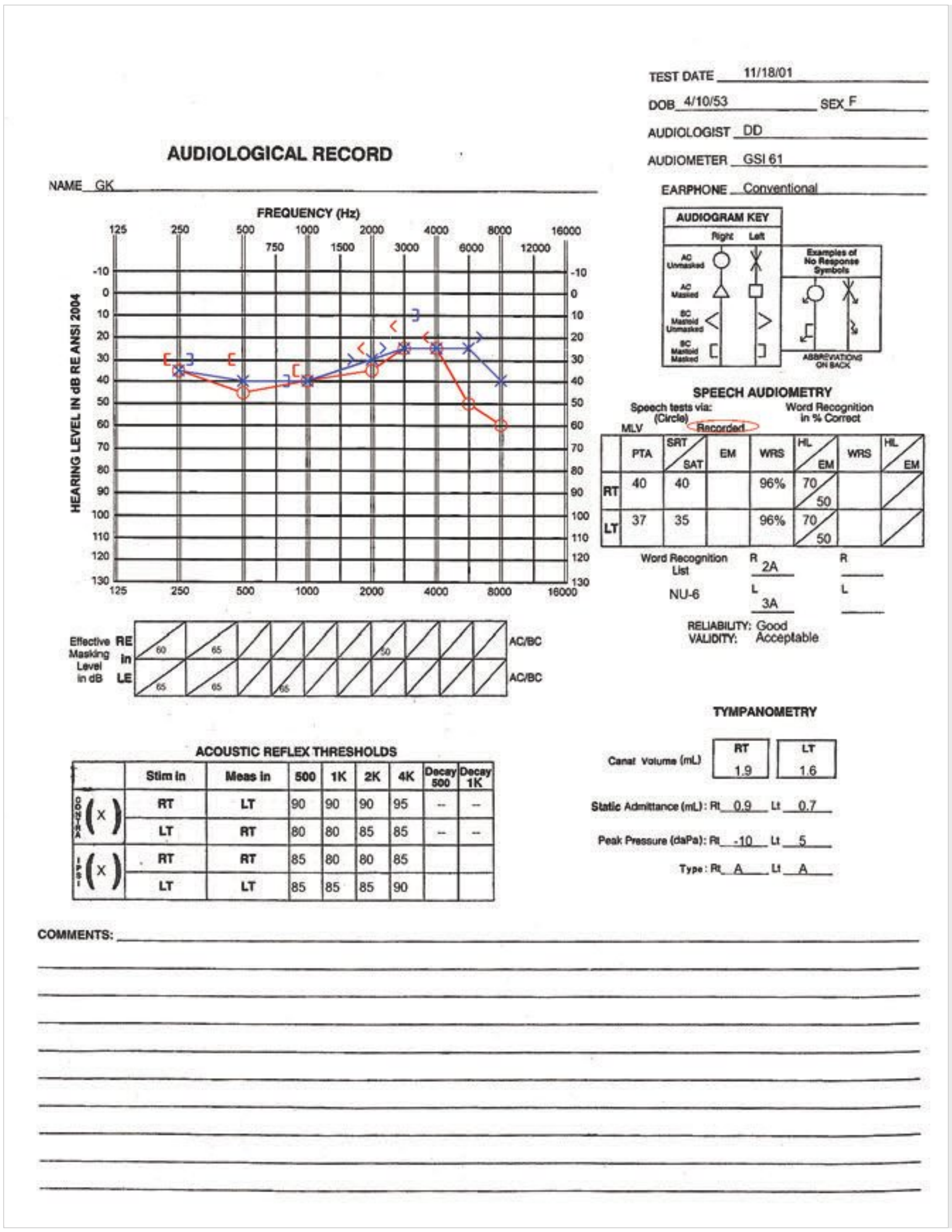
Judy L. Peterein

The patient in this case report will be referred to as RA. The case outlines the diagnosis and treatment of a patient with a right acoustic tumor, including long-term treatment effects and audiological rehabilitation.

11.1 Clinical History and Description

RA was originally seen in the audiology clinic when she was 51 years of age. She reported some degree of hearing loss for several years. She also reported bilateral tinnitus. Her mother had hearing loss, which prompted RA to schedule an evaluation. Her otologic history was otherwise negative. The initial audiogram revealed a mild to moderate bilateral sensorineural hearing loss through 2000 Hz rising to a slight hearing loss at 3000 and 4000 Hz. Pure-tone thresholds for the left ear revealed a slight loss at 6000 Hz and a mild loss at 8000 Hz. Pure-tone thresholds for the right ear revealed a moderate loss at 6000 Hz and a moderately severe loss at 8000 Hz. Speech recognition thresholds (SRTs) revealed a mild loss in the ability to receive speech and word recognition scores (WRSs) were within normal limits bilaterally. Tympanograms were within normal limits bilaterally. Ipsilateral and contralateral acoustic reflex thresholds (ARTs), as well as acoustic reflex decay measures, were also within normal limits bilaterally (Fig. 11.1).

RA was also seen by an otologist. Based on her history, physical examination, and audiological test results, the otologist recommended she return for repeat audiological testing in 2 years. RA was seen for a follow-up audiological examination 3 years later, where she reported progression of hearing loss in the right ear. In addition to the progression of hearing loss in her right ear she indicated slight imbalance but no vertigo. She also reported a recent sinus infection. Repeat audiological examination revealed pure-tone thresholds in the left ear essentially unchanged from 250 to 4000 Hz; however, there was a 20 dB HL



AUDIOLOGICAL RECORD

NAME GK

TEST DATE 11/18/01

DOB 4/10/53 SEX F

AUDIOLOGIST DD

AUDIOMETER GSI 61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT	40	40		96%	70/50		
LT	37	35		96%	70/50		

Word Recognition List NU-6

R 2A R

L 3A L

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	90	90	90	95	--	--
	LT	RT	80	80	85	85	--	--
IPSI (X)	RT	RT	85	80	80	85		
	LT	LT	85	85	85	90		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.9	1.6

Static Admittance (mL): Rt 0.9 Lt 0.7

Peak Pressure (daPa): Rt -10 Lt 5

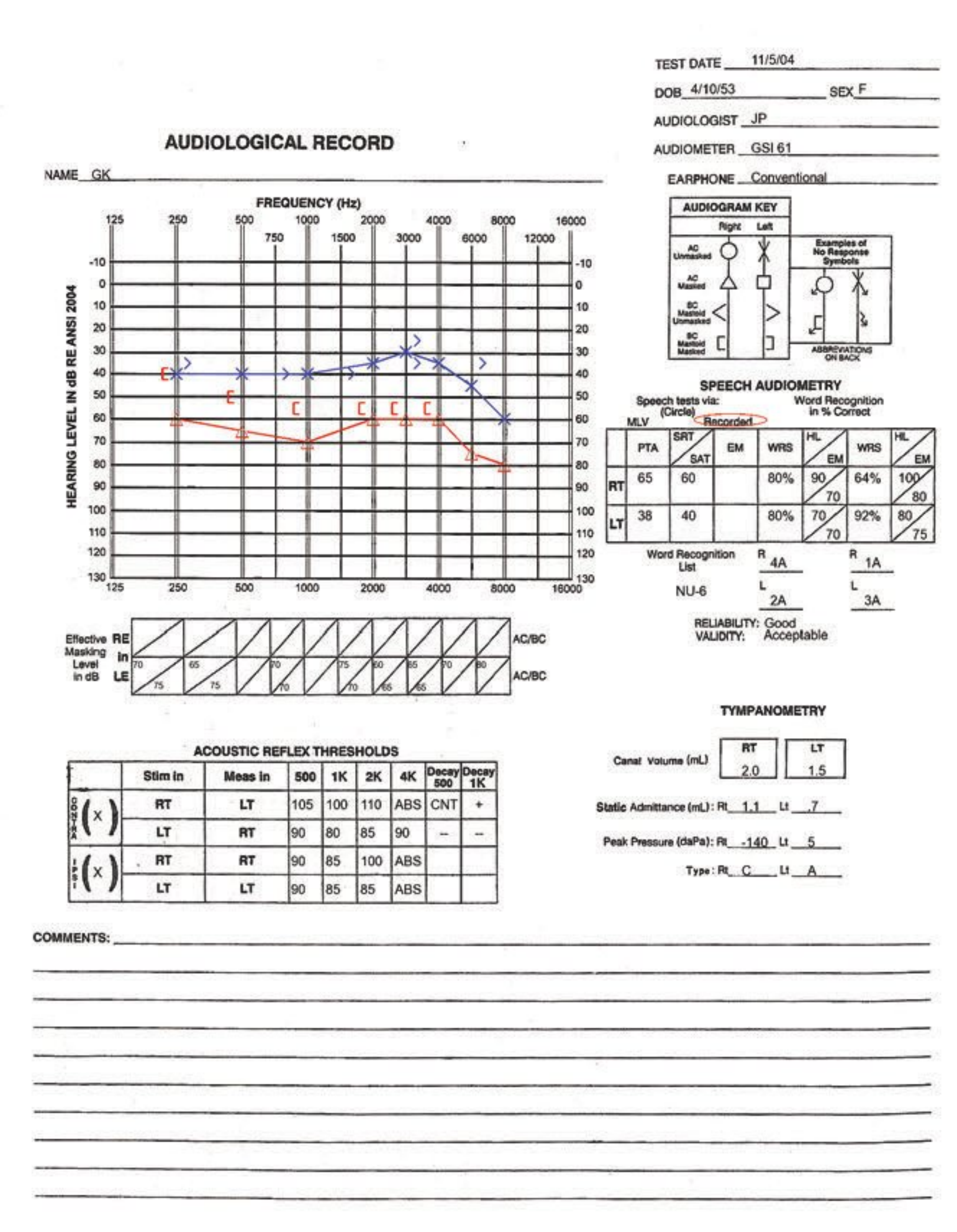
Type: Rt A Lt A

COMMENTS:

Fig. 11.1 Initial audiological examination (November 18, 2001).

decrease at 6000 and 8000 Hz. Pure-tone thresholds for the right ear revealed a significant decrease in air conduction thresholds at all test frequencies. A slight (15–20 dB HL) conductive component was noted at 250 to 1000 Hz. The WRS for the left ear remained within normal limits. The WRS for the right ear was slightly impaired (80%) when words were presented at 90 dB HL, with a moderate impairment (64%) noted when the presentation level was increased to 100 dB HL. Although WRS was reduced with increased presentation level, rollover was not significant. Tympanometry for the right ear revealed normal amplitude with middle ear pressure of − 140 daPa. Slight air–bone gaps for the right ear at 250 to 1000 Hz may have been due to negative middle ear pressure. The tympanogram for the left ear was within normal limits. Right contralateral ARTs were elevated at 500 and 2000 Hz, with positive acoustic reflex decay noted at 1000 Hz stimulating the right ear (□ Fig. 11.2). Due to the progression of hearing loss in the right ear, the otologist recommended a magnetic resonance imaging (MRI) scan. The MRI scan revealed a right enhancing cerebellopontine angle lesion extending into the right internal auditory canal consistent with a probable vestibular schwannoma, also referred to as an acoustic tumor. The lesion measured 1.4 × 0.8 cm—a small acoustic tumor.

RA was referred to the Department of Neurological Surgery for consultation regarding the small right acoustic tumor. RA was provided three options with regard to treatment of the tumor. First, serial MRI scans could be performed to monitor tumor size. Watchful waiting is an option for patients with small tumors. Second, the tumor could be removed surgically through either a translabyrinthine or suboccipital approach. Third, Gamma Knife radiation treatment could be used. RA chose to wait and consider her options and was seen by the neurosurgeon 5 months later. The MRI scan was repeated with tumor measurement described as 1.7 × 0.9 × 1.0 cm. The tumor was reported as just slightly increased in size as compared with the previous study. Audiological examination was also repeated. Test results for the left ear were unchanged when compared with the most recent examination. Test results for the right ear revealed a decrease in pure-tone thresholds of 15 dB HL at 500 Hz, 35 dB HL at 6000 Hz, and 10 dB HL at 8000 Hz. The WRS for the right ear decreased to poor (56%) with the presentation level at 90 dB HL and increased to


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

TEST DATE 11/5/04
DOB 4/10/53 SEX F
AUDIOLOGIST JP
AUDIOMETER GSI 61
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME GK

Effective Masking Level in dB

RE in											AC/BC
LE	70 75	65 75		70 70		75 70	60 65	65 65	70	80	AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT	65	60		80%	90/70	64%	100/80
LT	38	40		80%	70/70	92%	80/75

Word Recognition List NU-6 R 4A L 2A R 1A L 3A

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	2.0	1.5

Static Admittance (mL): Rt 1.1 Lt .7
Peak Pressure (daPa): Rt -140 Lt 5
Type: Rt C Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	105	100	110	ABS	CNT	+
	LT	RT	90	80	85	90	–	–
IPSI (X)	RT	RT	90	85	100	ABS		
	LT	LT	90	85	85	ABS		

COMMENTS:

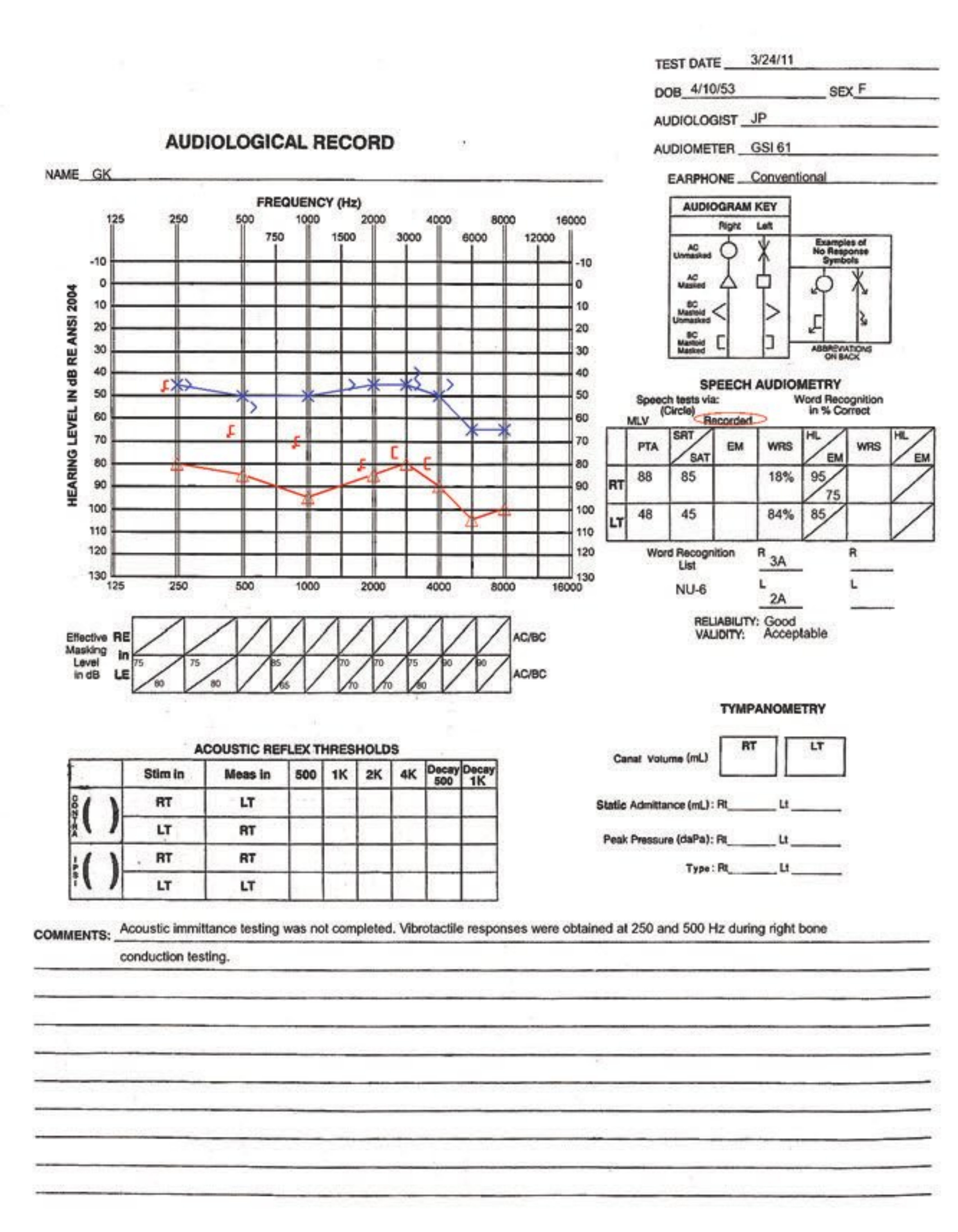
Fig. 11.2 Audiological examination completed 3 years after the initial audiogram (November 5, 2005). RA reported progression of hearing loss in her right ear and slight imbalance.

moderately impaired (70%) when the presentation level was increased to 100 dB HL. Middle ear pressure was normal, and the right tympanogram was within normal limits. RA decided to proceed with stereotactic radiosurgery using the Gamma Knife (Elekta, Stockholm, Sweden). She underwent Gamma Knife treatment approximately 2 weeks later.

RA was seen for follow-up by the neurosurgeon, the otologist, and the audiologist at regular intervals following Gamma Knife treatment. Annual MRI scans and annual audiograms were recommended and completed. An MRI scan completed approximately 1 year following Gamma Knife treatment revealed a slight decrease in tumor dimension suggesting some interval regression in response to Gamma Knife treatment. Audiological examination completed approximately 7 months following Gamma Knife treatment revealed additional decrease in pure-tone thresholds in the right ear of 10 dB HL at 250 Hz, 15 dB HL at 1000, 3000, 4000, and 8000 Hz and 25 dB HL at 2000 Hz. The WRS for the right ear remained moderately impaired (64%). Of note was that all right ipsilateral and contralateral ARTs were now absent. Audiometric findings for the left ear were stable. Subsequent MRI scans completed over the next 6 years revealed the size of the acoustic tumor to be stable. Serial audiograms, however, completed over the next 6 years revealed additional decrease in the pure-tone thresholds and WRS in the right ear. The most recent audiogram completed 6 years following Gamma Knife treatment revealed a severe to profound sensorineural hearing loss in the right ear with very poor (18%) word recognition ability. Pure-tone test results for the left ear remained stable. The left WRS was slightly impaired (84%) (Fig. 11.3).

11.2 Audiological Testing

Conventional audiological examination and acoustic immittance testing were completed at RA's initial evaluation as well as at her follow-up appointment 3 years later. Test results from the two appointments are shown in Fig. 11.1 and Fig. 11.2. Additional audiological examinations were completed prior to Gamma Knife treatment as well as annually. The most recent audiological examination was completed approximately 6 years following Gamma Knife treatment and is shown in Fig. 11.3.


FREQUENCY (Hz)
125 250 500 1000 2000 4000 8000 16000
750 1500 3000 6000 12000
HEARING LEVEL IN dB RE ANSI 2004
-10 0 10 20 30 40 50 60 70 80 90 100 110 120 130
AUDIOGRAM KEY
Right Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

AUDIOLOGICAL RECORD

NAME GK

TEST DATE 3/24/11

DOB 4/10/53 SEX F

AUDIOLOGIST JP

AUDIOMETER GSI 61

EARPHONE Conventional

Effective Masking Level in dB

RE in											AC/BC
LE	75 / 80	75 / 80		85 / 65		70 / 70	70 / 70	75 / 80	90	90	AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	88	85		18%	95 / 75		
LT	48	45		84%	85		

Word Recognition List R 3A R

NU-6 L 2A L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt_____ Lt_____

Peak Pressure (daPa): Rt_____ Lt_____

Type: Rt_____ Lt_____

COMMENTS: Acoustic immittance testing was not completed. Vibrotactile responses were obtained at 250 and 500 Hz during right bone conduction testing.

Fig. 11.3 Audiological examination completed 6 years following Gamma Knife treatment (March 24, 2011). Additional progression of hearing loss in the right ear including decrease in pure-tone thresholds and word recognition scores is noted.

11.3 Questions to the Reader

1. What initial audiological test results were consistent with the patient's final diagnosis?
2. What additional audiological tests could have been completed to further delineate hearing loss site of lesion?
3. Were the patient's initial symptoms consistent with the diagnosis of a right acoustic tumor?
4. How would you explain the continued progression of sensorineural hearing loss in the right ear following Gamma Knife treatment?

11.4 Discussion of Questions to the Reader

1. What initial audiological test results were consistent with the patient's final diagnosis?
   Significant pure-tone asymmetry was present at two consecutive frequencies (6000 and 8000 Hz) at the initial audiogram. Hearing loss asymmetry is a hallmark of retrocochlear hearing loss site of lesion. RA also had bilateral cochlear hearing loss, and therefore the hearing loss in the right ear would be considered mixed cochlear/retrocochlear. Bilateral tinnitus may have been due to cochlear dysfunction bilaterally; therefore, the possible retrocochlear red flag of unilateral tinnitus was not apparent in this case. In addition, WRS was normal bilaterally, possibly due to the small tumor size.
2. What additional audiological tests could have been completed to further delineate hearing loss site of lesion?
   Standard auditory brainstem response (ABR) testing may have been helpful in early identification of retrocochlear dysfunction. The standard ABR may have been normal due to the small tumor size, but in approximately 60 to 70%of small acoustic tumors, abnormalities in the standard ABR are noted. Referral for ABR testing due to pure-tone asymmetry may have resulted in earlier identification of the right acoustic tumor.
3. Were the patient's initial symptoms consistent with the diagnosis of a right acoustic tumor?
   Initial patient symptoms did not strongly suggest the presence of a right acoustic tumor. RA reported general bilateral hearing loss as well as bilateral tinnitus. Family history of hearing loss as well as hearing loss configuration (i.e., rising configuration) suggested some genetic hearing loss component. Unilateral symptoms were not reported at the initial evaluation. As discussed previously, the presence of asymmetry suggested possible retrocochlear involvement. Both cochlear and retrocochlear dysfunction were present in patient RA Conventional audiological examination cannot rule out retrocochlear dysfunction.
4. How would you explain the continued progression of sensorineural hearing loss in the right ear following Gamma Knife treatment?
   In general, progression of hearing loss is expected with increased acoustic tumor size. This, however, is not always true. It is possible to have a large cerebellopontine angle tumor with normal hearing test results using conventional audiometric procedures. Serial MRI scans revealed the right acoustic tumor was slightly smaller following Gamma Knife treatment. Thus hearing loss progression due to tumor growth would not be suggested in this case. It is possible that additional anatomical or physiological changes at the tumor–nerve interface may have occurred that were not identifiable via MRI scans. The effects of radiation from Gamma Knife treatment, however, are strongly suggested with regard to ongoing progression of hearing loss in RA's right ear. Current research is not in agreement regarding the relationship between radiation therapy, including Gamma Knife and possible long-term effects on hearing. Various case reviews in these studies have suggested (1) greater decrease of hearing following radiation treatment than in groups of nonirradiated patients, (2) equivalent hearing loss progression following radiation treatment as compared with patients who did not receive radiation, and (3) less hearing loss in groups of patients receiving radiation compared with patients who did not receive radiation. Most investigators agree that a lower mean cochlear radiation dose (<4 Gy) is an important factor related to hearing preservation. Additional controlled studies are necessary to determine long-term effects of radiation treatment, including Gamma Knife, on progression of hearing loss.

11.5 Diagnosis and Recommended Treatment

Medical diagnosis and treatment for patient RA have been discussed. Audiological treatment is described in the following section. RA was fit with a hearing aid for her left ear prior to Gamma Knife treatment. Hearing aid options including the possibility of a left bilateral contralateral routing of the signal (BICROS) hearing aid were discussed, but RA elected to be fit only with a hearing aid in her left ear. Initial hearing aid fitting was completed with a midlevel digital behind-the-ear (BTE) hearing instrument and a custom Lucite skeleton earmold. Real ear measures were performed using a Frye 6500 real ear analyzer (Frye Electronics, Inc., Tigard, OR) to verify that the hearing aid settings provided gain that met the National Acoustic Laboratories' non-linear prescription, version 1 (NAL-NL1) prescriptive target for an input level of 65 dB (sound pressure level). RA successfully wore the left hearing aid for the next 5 years. After 5 years, RA replaced the original left digital hearing aid with a significantly advanced 16-channel digital BTE hearing aid with automatic directionality. Once again, real ear measures were used to verify hearing aid performance. In addition to the real ear testing completed with the original hearing aid, input levels of 50 and 80 dB SPL were also used to verify the nonlinear performance of the new hearing aid. Directional microphone performance was then verified using real ear measures. RA noticed significant improvement in daily communication with the new left digital hearing aid. She wore this hearing aid for approximately 2 years before she decided to add a Phonak CROSLINK system (Phonak U.S., Warrenville, IL) to the right ear. She has been successfully wearing the wireless digital left BTE BICROS hearing aid since that time.

11.6 Outcome

Currently Gamma Knife treatment has been successful in arresting acoustic tumor growth for RA. She successfully wears a wireless digital left BTE BICROS hearing aid.

11.7 Key Points

1. Significant hearing loss asymmetry should be evaluated further at the time of the initial examination.
2. Treatment options for vestibular schwannoma/acoustic tumor include serial MRI scans to monitor tumor growth, surgical tumor removal, and radiation treatment.
3. Although additional systematic study is needed, evidence suggests that radiation treatment for acoustic tumors results in the progression of hearing loss long term.
4. The use of contralateral routing of the signal (CROS) and BICROS hearing aid technology for patients following acoustic tumor treatment can provide significant benefit.

Suggested Reading

Gordon ML, Cohen NL. Efficacy of auditory brainstem response as a screening test for small acoustic neuromas. Am J Otol 1995; 16: 136–139

Maniakas A, Saliba I. Conservative management versus stereotactic radiation for vestibular schwannomas: a meta-analysis of patients with more than 5 years' follow-up. Otol Neurotol 2012; 33: 230–238

Rasmussen R, Claesson M, Stangerup SE et al. Fractionated stereotactic radiotherapy of vestibular schwannomas accelerates hearing loss. Int J Radiat Oncol Biol Phys 2012; 83: e607–e611

Roos DE, Potter AE, Zacest AC. Hearing preservation after low dose linac radiosurgery for acoustic neuroma depends on initial hearing and time. Radiother Oncol 2011; 101: 420–424

Yomo S, Carron R, Thomassin JM, Roche PH, Régis J. Longitudinal analysis of hearing before and after radiosurgery for vestibular schwannoma. J Neurosurg 2012; 117: 877–885

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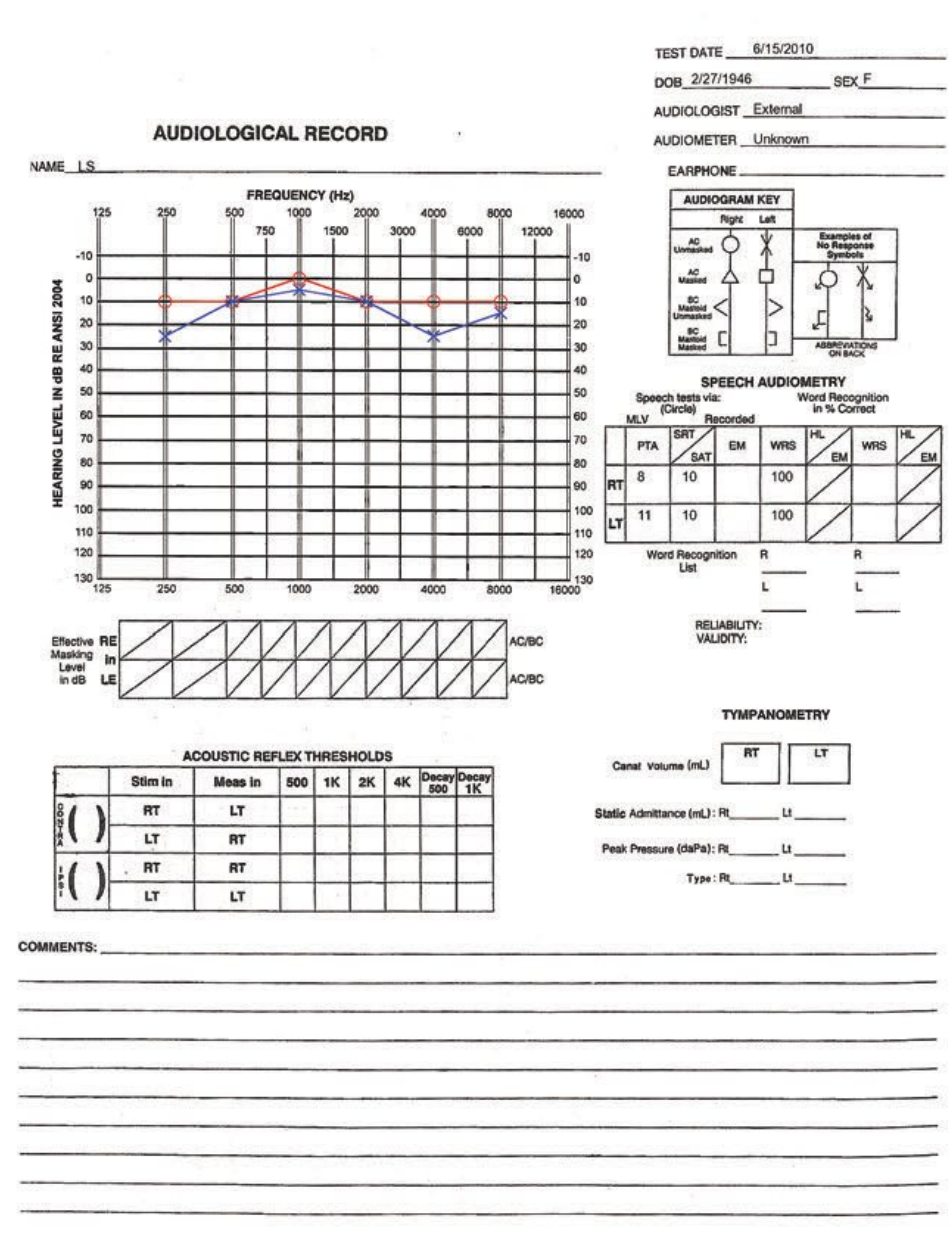
12 Vestibular Schwannoma

Gerald R. Popelka

Audiological measures in cases of vestibular schwannoma play an important role in detecting such cases and monitoring auditory and vestibular changes in relation to intervention. The audiologist also plays an important role in providing counseling and appropriate devices for any resultant hearing loss.

12.1 Clinical History and Description

LS is a 63-year-old woman originally seen in an otolaryngology outpatient clinic with a primary report of aural fullness in the left ear. Because she had an audiological examination from another institution within the past year and had no report of hearing difficulties, LS was automatically scheduled only for an otology appointment rather than combined audiology and otology appointments. The audiogram from the other institution (Fig. 12.1) reported normal hearing by air conduction with a four-frequency (500, 1000, 2000, and 3000 Hz) pure-tone average (PTA) of 8 dB HL for the right ear and 11 dB HL for the left ear. There were no otoscopy results, tympanograms, bone conduction results, or acoustic reflex thresholds (ARTs) reported. Speech recognition thresholds (SRTs) were 10 dB HL bilaterally, within normal limits and consistent with the PTAs. Word recognition scores (WRSs) in quiet were 100%at 55 dB HL bilaterally. The only remarkable finding was the slightly elevated thresholds at two nonadjacent frequencies (4000 Hz and 8000 Hz) in the left ear compared with the right ear. The slight asymmetry (15 dB) at 250 Hz may not normally be of concern because this finding is easily consistent with a very mild temporary conductive component associated with common upper respiratory infections, allergies, excess cerumen, and the



TEST DATE 6/15/2010

DOB 2/27/1946 SEX F

AUDIOLOGIST External

AUDIOMETER Unknown

EARPHONE

AUDIOLOGICAL RECORD

NAME LS

Effective Masking Level in dB RE in AC/BC
LE AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT	8	10		100			
LT	11	10		100			

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt ____ Lt ____

Peak Pressure (daPa): Rt ____ Lt ____

Type: Rt ____ Lt ____

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 12.1 Initial audiological results obtained from an external institution prior to the initial otologic evaluation and prior to a magnetic resonance imaging scan. Note the threshold at 3000 Hz was not measured, a four-frequency pure-tone average can be computed by interpolating the value using thresholds at 2000 Hz and 4000 Hz.

complaint of aural fullness. Otoscopy results, bone conduction thresholds, ARTs, and tympanograms would have contributed to understanding this minor threshold elevation but were not provided. The type of earphone used would also have been beneficial because a minor elevation of air conduction threshold at this lower frequency is consistent with a slight acoustic leak commonly seen with supra-aural earphones, but not with insert earphones. The slight asymmetry (15 dB) at 4000 Hz also may not be of concern because a slightly elevated air conduction threshold at 4000 Hz is consistent with noise exposure, but no noise exposure was provided.

During the otology exam, LS reported a history of an occipital osteoma, a slowly growing benign tumor of the bone in the skull bone that was repaired when LS was a child. She also reported occasional and very mild tinnitus in her left ear and a very minimal history of vertigo and disequilibrium and no significant noise exposure. The otology physical exam showed normal external ear canals, tympanic membranes, and middle ear spaces bilaterally under binocular microscopy. Weber and Rinne tests at 512 Hz did not reveal any asymmetry nor a conductive component. The remainder of the otology physical examination of the oral cavity, oropharynx, anterior nasal cavities, and neck showed no abnormal findings. The neurological portion of the exam did not reveal any abnormalities of the trigeminal nerve, the corneal reflexes, nor the cerebellar measures. There were no other obvious neurological signs such as facial twitching, facial paresthesias, or facial weakness, but there was a minor complaint of headaches. There were no grossly observable extraocular eye movements, nor grossly observable abnormal nystagmus. The Romberg test was normal. Gait observations including tandem gait with eyes open and eyes closed were also normal. The ability to step in place with eyes closed (Fukuda test) was also normal.

Though no obvious single pathology was suggested by these results, the overall pattern initially raised several concerns, including cochlear hydrops, because of the slight low-frequency asymmetry, lateral semicircular canal dehiscence that can show a conductive component in the low frequencies, but there were no bone conduction thresholds indicated, and some concern about headaches and the previous skull tumor. For all these reasons, only partially related to the asymmetrical and missing audiometric results, LS received a magnetic resonance imaging (MRI) scan with gadolinium contrast a few days later. The MRI scan indicated evidence of the previously repaired occipital osteoma. The scan also revealed a vestibular schwannoma, also called an acoustic neuroma. This benign, space-occupying tumor, 13 mm in its greatest dimension, was located entirely in the left internal auditory canal. Because the vestibular schwannoma was definitively diagnosed based on MRI scan, no additional audiological measures such as vestibular evaluation or auditory evoked responses were necessary.

A vestibular schwannoma, often called an acoustic neuroma, is a benign, slowly growing, intracranial tumor that originates from the Schwann cells which form the myelin sheath that covers the vestibular portion of the eighth cranial nerve (CN VIII) and that also includes the auditory nerve from the cochlea. Because this nerve and the seventh cranial nerve (CN VII) course within the tight confines of the internal auditory canal where most of these tumors originate, auditory, vestibular, and facial symptoms may be expected. The prevalence is about 1/100,000 worldwide with about 3000 new cases diagnosed annually in the United States. The highest incidence occurs between 50 and 60 years of age and is distributed equally between males and females. Though very rare in the general population, these tumors represent a significant percentage of all intracranial tumors in adults.

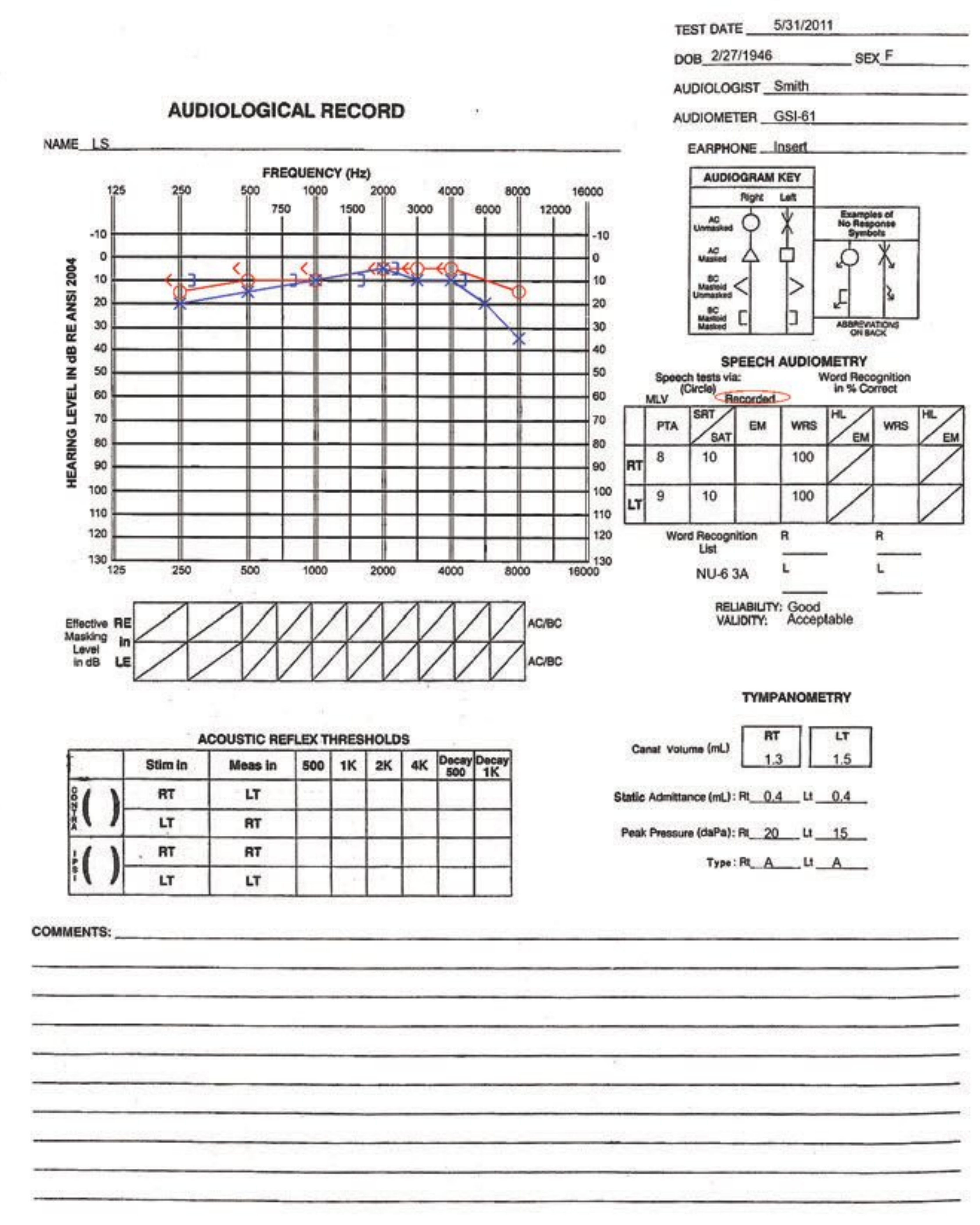
Initial auditory symptoms include unilateral sensorineural hearing loss, tinnitus, a sense of aural fullness, and difficulty recognizing speech. Initial vestibular symptoms include balance disorders, changes in gait, and vertigo. Initial facial symptoms occur less often but include abnormal facial muscle function. In cases of larger tumors that extend out of the internal auditory canal toward the brainstem, other cranial nerves can be affected, including the fifth cranial nerve (CN V), which can result in sensation changes on the face. The glossopharyngeal nerve (CN IX) and vagus nerve (CN X) are not typically involved, but their involvement may lead to abnormal gag or swallowing reflexes. Larger tumors may also increase the intracranial pressure, resulting in other symptoms such as headaches and nausea.

This case report is illustrative of the role of asymmetrical hearing in the detection of retrocochlear disease, the importance of monitoring hearing in relation to hearing preservation treatment options, and consideration of devices used for unilateral hearing loss (UHL).

There are three common interventions for treating vestibular schwannoma. One common approach is to do no intervention and monitor the tumor growth and development of symptoms because these tumors often do not grow very rapidly. The second common intervention is to prevent further growth and possible shrinkage of the tumor with stereotactic radiation (Gamma Knife, Elekta, Stockholm, Sweden; or CyberKnife, Accuray, Sunnyvale, CA) an approach that does not require surgery. The third common intervention is surgical removal of the tumor through a craniotomy using microsurgical techniques. After discussion with the patient of the advantages and disadvantages of the three possible interventions, LS decided on no intervention other than monitoring. This is a very common and appropriate intervention given the mildness of the symptoms and the fact that the tumor was entirely within the internal auditory canal and with no information on rate of tumor growth, which is likely slow. With this choice, the recommendation was for a complete audiological examination and otology evaluation in 1 year, or sooner in the event of noticeable change.

12.2 Audiological Testing

A more comprehensive audiological examination (Fig. 12.2) obtained with insert earphones 12 months after the initial external audiogram again reported normal hearing sensitivity by air conduction with a PTA of 8 dB HL for the right ear and 9 dB HL for the left ear. The otoscopy results revealed clear external ear canals and normal tympanic membranes bilaterally. Bone conduction thresholds were equivalent to the air conduction thresholds at all frequencies, including 250 Hz. The tympanometric results, including equivalent volume, peak



AUDIOLOGICAL RECORD

NAME LS

TEST DATE 5/31/2011
DOB 2/27/1946 SEX F
AUDIOLOGIST Smith
AUDIOMETER GSI-61
EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	8	10		100			
LT	9	10		100			

Word Recognition List NU-6 3A
R
L
R
L

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB
RE AC/BC
LE AC/BC

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.3	1.5

Static Admittance (mL): Rt 0.4 Lt 0.4
Peak Pressure (daPa): Rt 20 Lt 15
Type: Rt A Lt A

COMMENTS:

Fig. 12.2 Audiological results 12 months later and obtained just prior to stereotactic radiation for a vestibular schwannoma in the left ear.

pressure, and static admittance values, all were in the normal range bilaterally. Ipsilateral ARTs at three frequencies (500, 1000, and 2000 Hz) were in the normal range (85 to 90 dB HL) for the right ear and absent at the maximum levels (>100 to 105 dB HL) for the left ear. The SRT of 10 dB HL in each ear was consistent with the PTA, and the WRS in quiet was 100% at 50 dB HL in each ear. No new auditory or vestibular symptoms were reported by the patient.

There are several remarkable findings in these audiometric results (Fig. 12.2) compared with the initial audiometric results (Fig. 12.1). First, there was no external ear canal, tympanic membrane, or middle ear involvement based on the equivalence of the air and bone conduction thresholds, the normal otoscopy results, the normal tympanometry results, and the elimination of any earphone acoustic leak with the use of an insert earphone. These results strongly suggested that the initial complaint of aural fullness was not associated with external or middle ear disorders or cochlear hydrops. Second, with no conductive system abnormalities on either side, the asymmetry in the ipsilateral ARTs suggests that there was significant involvement of the sensorineural system on the left side. Further, because the left hearing thresholds were in the normal hearing range, the retrocochlear system should be of concern. Third, the lack of an asymmetry of thresholds at 250 Hz suggests that the previous asymmetry at this frequency could have been related either to a temporary conductive or earphone issue, as suggested earlier, or to actual but slightly variable thresholds in the sensorineural system. Fourth, the lack of an asymmetry of thresholds at 4000 Hz suggests that the previous slight elevation in threshold at this frequency was related to retrocochlear issues because noise exposure was not a factor, and the possible conductive concern raised when interpreting the initial audiogram does not usually reveal itself at this higher frequency. Finally, the largest asymmetry for either audiogram (20 dB HL) was seen at 8000 Hz in the second audiogram even though there was no asymmetry at this frequency in the first audiogram.

The repeat MRI scan revealed that the vestibular schwannoma had grown larger. For this reason, LS elected to begin stereotactic radiation intervention (CyberKnife) and received four treatments over 4 days and a recommendation for another comprehensive audiological examination in six months.

12.3 Questions to the Reader

1. Why was the pure-tone audiogram asymmetry the only audiological indication of this problem initially?
2. What constitutes asymmetry?
3. Do subsequent audiological examinations need to be tailored to the various specific interventions that may include no intervention, stereotactic radiation, or surgery?
4. Can electronic medical record (EMR) systems help in providing a decision concerning the quantification of asymmetry that relates to the detection and monitoring of symptoms associated with vestibular schwannoma?
5. Should specific audiological interventions be considered in cases of vestibular schwannoma?

12.4 Discussion of Questions to the Reader

1. Why was the pure-tone audiogram asymmetry the only audiological indication of this problem initially?
   The correlation between certain vestibular schwannoma characteristics, such as the size of the tumor, and hearing sensitivity is not high. The vestibular schwannoma arises from the vestibular branch of the eighth cranial nerve (CN VIII), suggesting that vestibular signs should always be present. Because most of these tumors grow slowly, however, the patient often adapts to the compromised vestibular system function with the use of the remaining normally functioning vestibular system, the visual system, and the proprioceptive systems. It is not unusual for a patient with a small vestibular schwannoma not to complain of balance or mobility problems or vertigo.
2. What constitutes asymmetry?
   A specific criterion of what constitutes an asymmetry has never been widely adopted. Because of this, it is not surprising that even experienced clinicians may classify the same audiogram differently or that a large percentage of all audiograms may be categorized as asymmetric even if there is no conductive component. A decision for making a medical referral is affected more by the clinical setting, the clinical experience of the audiologist, and factors other than hearing sensitivity.

   Many criteria for what constitutes asymmetry have been proposed, but none have been standardized. These include an asymmetry in thresholds of specified decibel differences at multiple individual frequencies, a single frequency, or in the average of the thresholds at defined frequencies such as the PTA.

   If an asymmetry with small differences is considered, a variety of factors unrelated to retrocochlear pathology will influence the result. These include the normal variability of threshold changes associated with variable external ear canal, tympanic membrane and middle ear conditions, threshold measurement protocols, and earphone type. There is less concern about these factors with large asymmetries. Finally, pure-tone hearing sensitivity is not the only measure of asymmetry. Acoustic reflex thresholds, interpeak and interaural peak latencies in auditory evoked brainstem responses and other measures may also fulfill a definition of asymmetry.
3. Do subsequent audiological examinations need to be tailored to the various specific interventions that may include monitoring, stereotactic radiation, or surgery?
   Audiology plays an important role in providing information for choosing specific intervention and in monitoring auditory and vestibular function from that point on. All three interventions have a hearing preservation purpose, and research does not support one of these interventions as having markedly better success at hearing preservation compared with the others, so having accurate hearing measures can influence the choice of intervention. After an intervention approach has been decided and implemented, continued accurate measures of hearing remain important. Even if no intervention is implemented, frequent audiological examinations are needed to detect any additional changes in auditory or vestibular function. Any observed change in hearing or vestibular function may alter the intervention strategy from no intervention to one of the two other intervention options. If either of the remaining interventions, radiation or surgery, is implemented, any observed change in hearing or vestibular function may result in changing the selected strategy. These include consideration of additional sessions of radiation, termination of radiation and deciding on surgery, or in some cases when surgery has not removed all of the tumor, the addition of radiation after surgery. In all cases, accurate measures of hearing sensitivity are important for making these decisions, both for the patient and for the individuals providing the interventions.
4. Can EMR systems help in making a decision concerning the quantification of asymmetry that relates to the detection and monitoring of symptoms associated with vestibular schwannoma?
   A large majority of the audiological data that relate to the detection and monitoring of symptoms associated with vestibular schwannoma are amenable to quantification and analysis. These include the numeric values for the pure-tone thresholds, tympanometric values, patient age and certain binary information such as gender. Even more subjective information such as noise exposure history (e.g., none, mild, moderate) can be encoded into an EMR to allow the system to automatically perform numeric calculations and analysis. This has many advantages in that unrelated conductive components can be accounted for, arithmetic errors can be eliminated, missing data can be interpolated (e.g., a missing threshold at 3000 Hz can be computed from the thresholds at 2000 and 4000 Hz), and different asymmetry protocols can be calculated simultaneously. Paper-based systems, of which many remain, cannot do this. Unfortunately, even many of the newer EMR systems becoming increasingly available record the audiograms only as scanned images, rather than numerical values, thereby preventing any numerical analysis at all. A numerically based EMR system can automatically and accurately determine relevant asymmetries, including integration of non numeric characteristics such as noise history. This approach produces reliable results and can facilitate standardization. A calculation of the asymmetry that includes pure-tone air conduction thresholds at six frequencies, age, gender, and a simple rating of the

history of noise exposure can be determined with a calculation based on a regression analysis of a large number of cases. This analysis indicated that if the audiometric pattern in Fig. 12.1 was considered asymmetrical, 54% of the cases would likely have a vestibular schwannoma, with a false-positive rate of 10% If the audiometric pattern in Fig. 12.2 was considered asymmetrical, 62% of the cases would likely have a vestibular schwannoma, but with a false-positive rate of 18% Such analysis can improve decisions for referral and would likely be improved further if any minor conductive components were considered.

5. Should specific audiological interventions be considered in cases of vestibular schwannoma?
   The hearing and vestibular outcomes of all interventions of vestibular schwannoma can be quite variable. Any of the interventions can result in additional hearing problems ranging from minor changes in hearing sensitivity to additional changes in ability to hear speech in quiet or noise, to additional moderate changes in hearing that change the patient from a binaural listener to a monaural listener along with all of the changes in spatial hearing that are related to monaural hearing. One outcome can be the observation that the tumor is completely removed during surgery with essentially normal hearing preserved. After additional MRI scans and audiological examinations reveal no changes over a period of 2 to 5 years postsurgery, the frequency of these examinations can be decreased or even stopped. On the other hand, another outcome can be total loss of hearing in one ear resulting from the surgery, in which case the patient would immediately change from an essentially normal-hearing individual to one with a profound UHL Yet another outcome may be the onset of clinically significant age-related hearing loss that may be complicated by any additional hearing loss from the vestibular schwannoma in cases where there was no radiation or surgical intervention. Finally, there may also be vestibular consequences. In general, middle-aged or younger patients with a compromised vestibular system are likely to adapt quite well to the reduced vestibular function, whereas older patients are less so, making it necessary to monitor vestibular signs over time as well with a frequency of monitoring related to the initial age of the patient.
   Patients with a severe or profound UHL following the removal of a vestibular schwannoma or following stereotactic radiation are significantly disabled in a number of situations such as hearing sounds from the unaidable side, hearing in the presence of background noise (both in quiet and noisy surroundings), and localizing sounds. Patients with UHL experience significant difficulties in group discussions and listening situations such as environments with fixed sources of noise where it is difficult to compensate for the disorder by changing position. The perceived and actual hearing disorder can be substantial. Patients should be realistically counseled regarding the communication difficulties of potential UHL that may occur with all forms of medical intervention including monitoring. Should hearing difficulties occur after these interventions, the patients should be offered one of the many effective devices for UHL such as osseointegrated and implanted bone conduction devices, dental bone conduction devices, and possibly a contralateral routing of signals (CROS) hearing aid.

12.5 Additional Testing

Additional audiological testing could have included a comprehensive vestibular evaluation with videonystagmography (VNG) calorics and positional and tracking measures, vestibular evoked myogenic potentials (VEMPs) for semicircular canal dehiscence, and evoked auditory brainstem responses (ABRs) for retrocochlear disease. The definitive diagnosis of a vestibular schwannoma with imaging, however, made these additional audiological examinations unnecessary.

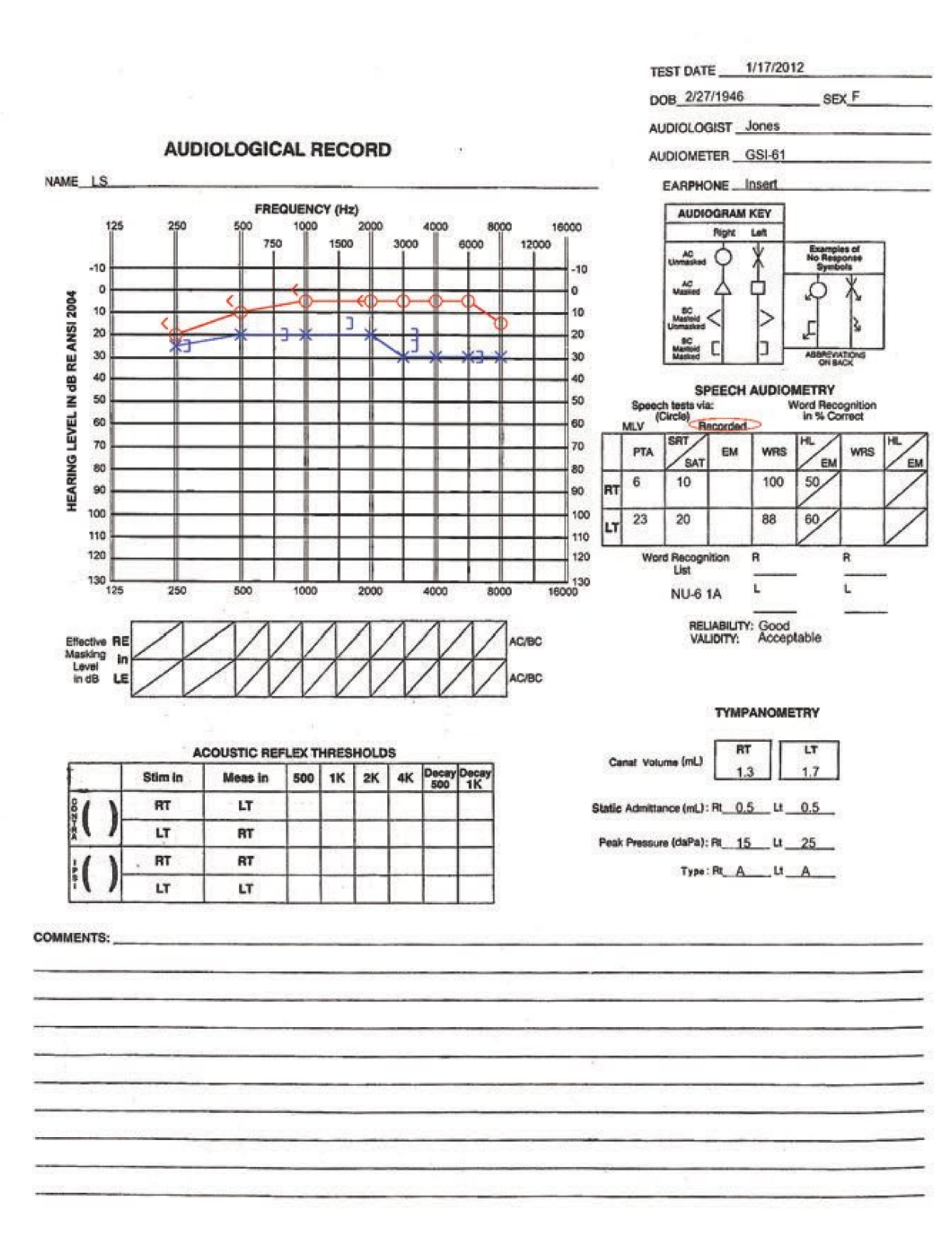
12.6 Diagnosis and Recommended Treatment

None of the likely beneficial advanced audiological examinations was necessary in this case because the definitive diagnosis of an intrcanalicular tumor, likely a vestibular schwannoma, was made with an MRI scan. The comprehensive audiological record and imaging were sufficient to determine the possible interventions. Realistic counseling concerning the possible changes in auditory and vestibular function was included as part of the treatment plan.

12.7 Outcome

Another audiological examination (Fig. 12.3) 6 months after the stereotactic radiation treatment revealed a slight decrease in hearing sensitivity and a more obvious asymmetry. This is not surprising in that stereotactic radiation often causes the tumor to swell initially and then shrink. Pure-tone audiometry again revealed essentially normal overall hearing by air conduction with a PTA of 6 dB HL for the right ear and 23 dB HL for the left ear. Even with this more obvious asymmetry, thresholds for frequencies below 3000 Hz were in the normal range (≤25 dB HL). The otoscopy results revealed clear external ear canals and normal tympanic membranes bilaterally. Bone conduction thresholds were equivalent to the air conduction thresholds at all frequencies, including 250 Hz and 4000 Hz. The tympanometric results, including equivalent volumes, peak pressures, and static admittance values, all were in the normal range bilaterally. If the audiometric pattern in Fig. 12.3 was considered asymmetrical, 62% of the cases would likely have a vestibular schwannoma with a false-positive rate of 18% Ipsilateral ARTs were not measured because they would not have added any useful information. The SRTs were consistent with the PTAs bilaterally. The WRSs in quiet were 100% at 50 dB HL for the right ear, but slightly decreased at 88% in the left ear at 60 dB HL Though this asymmetry existed, the patient had completely serviceable bilateral hearing, and no devices were considered.

Another audiological examination (Fig. 12.4) 7 months later and 13 months after the stereotactic radiation treatment revealed essentially the same results as the previous audiological examination. There remained a slight decrease in hearing sensitivity and a more obvious asymmetry. Pure-tone audiometry again revealed essentially normal overall hearing by air conduction with a PTA of 6 dB HL for the right ear and 20 dB HL for the left ear. The thresholds for frequencies to 4000 Hz were still in the normal range (≤25 dB HL). If the audiometric pattern in


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

AUDIOLOGICAL RECORD

NAME LS

TEST DATE 1/17/2012

DOB 2/27/1946 SEX F

AUDIOLOGIST Jones

AUDIOMETER GSI-61

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT	6	10		100	50		
LT	23	20		88	60		

Word Recognition List NU-6 1A R ____ L ____ R ____ L ____

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB												
RE in												AC/BC
LE												AC/BC

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.3	1.7

Static Admittance (mL): Rt 0.5 Lt 0.5

Peak Pressure (daPa): Rt 15 Lt 25

Type: Rt A Lt A

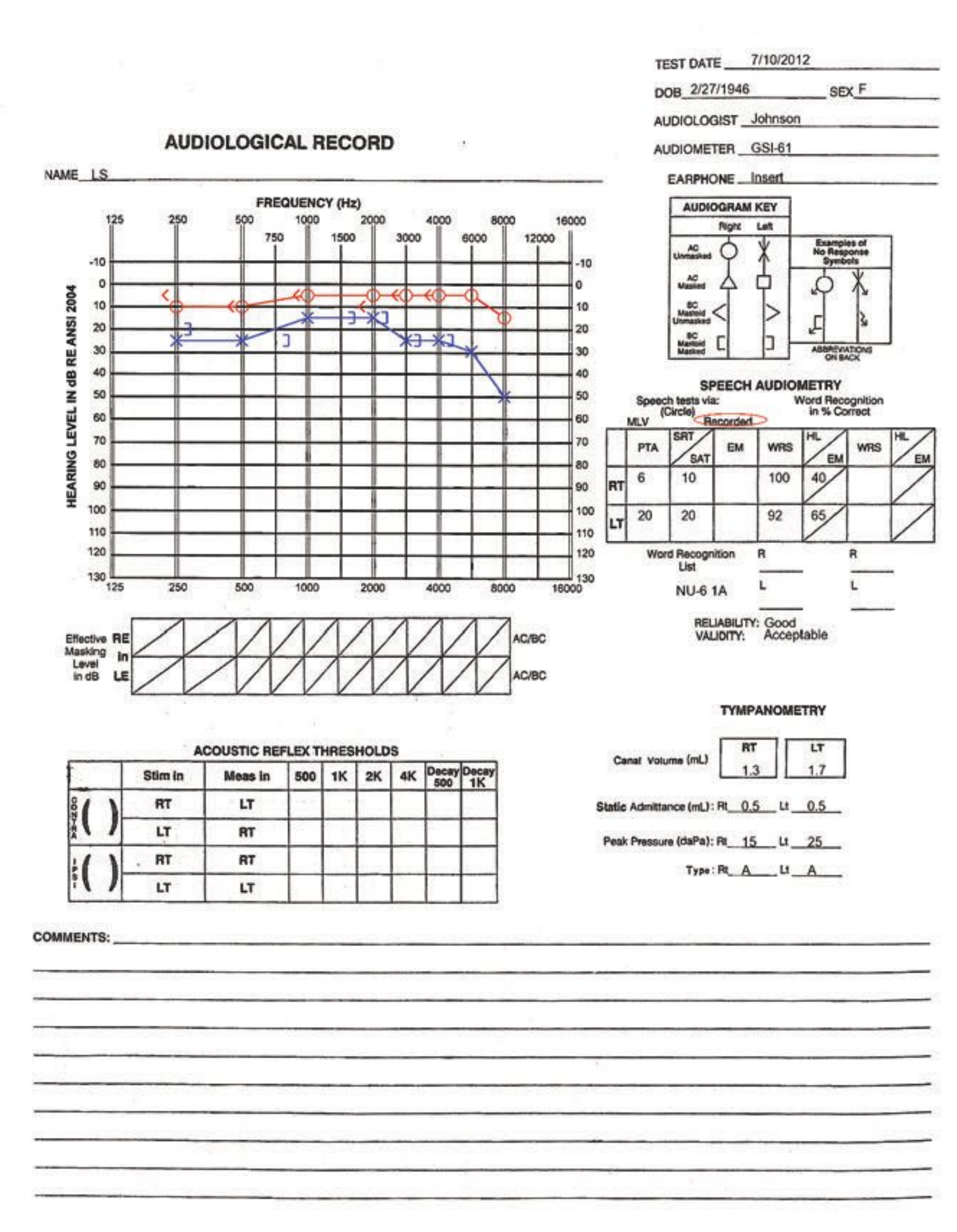
COMMENTS:

Fig. 12.3 Audiological results obtained 6 months after stereotactic radiation for a vestibular schwannoma in the left ear.

Fig. 12.4 was considered asymmetrical, 62% of the cases would likely have a vestibular schwannoma with a false-positive rate of 18%, but with a very low false-negative rate of < 1%. The otoscopy results revealed clear external ear canals and normal tympanic membranes bilaterally. Bone conduction thresholds were equivalent to the air conduction thresholds at all frequencies, including 250 Hz and 4000 Hz. The tympanometric results, including equivalent volumes, peak pressures, and static admittance values, all were in the normal range bilaterally. Ipsilateral ARTs again were not measured because they would not have added any useful information. The SRTs were consistent with the PTAs, and the WRSs in quiet were 100% at 40 dB HL for the right ear, still slightly decreased from normal but slightly higher compared with the previous audiological examination at 92% in the left ear at 65 dB HL. Though this asymmetry existed, LS had completely serviceable bilateral hearing and no devices were considered.

12.8 Key Points

1. Pure-tone audiometry can play an important role in detecting retrocochlear symptoms from a vestibular schwannoma.
2. Pure-tone audiometry can play an important role in monitoring any changes in hearing due to progression of the tumor, radiation therapy, or surgical intervention.
3. The audiologist can play an important role in proper counseling of the patient concerning possible changes in hearing function from intervention and providing one of several effective devices for UHL should no useable hearing remain in the affected ear.



AUDIOLOGICAL RECORD

NAME LS

TEST DATE 7/10/2012
DOB 2/27/1946 SEX F
AUDIOLOGIST Johnson
AUDIOMETER GSI-61
EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM	WRS	HL/EM
RT	6	10		100	40		
LT	20	20		92	65		

Word Recognition List NU-6 1A R L R L

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB RE in LE AC/BC AC/BC

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.3	1.7

Static Admittance (mL): Rt 0.5 Lt 0.5
Peak Pressure (daPa): Rt 15 Lt 25
Type: Rt A Lt A

COMMENTS:

Fig. 12.4 Audiometric results obtained 12 months after stereotactic radiation for a vestibular schwannoma in the left ear.

Suggested Reading

Gurgel RK, Popelka GR, Oghalai JS, Blevins NH, Chang KW, Jackler RK. Is it valid to calculate the 3-kilohertz threshold by averaging 2 and 4 kilohertz? Otolaryngol Head Neck Surg 2012; 147: 102–104

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13 Auditory Nerve Disorder: Structure versus Function

Brad A. Stach and James F. Jerger

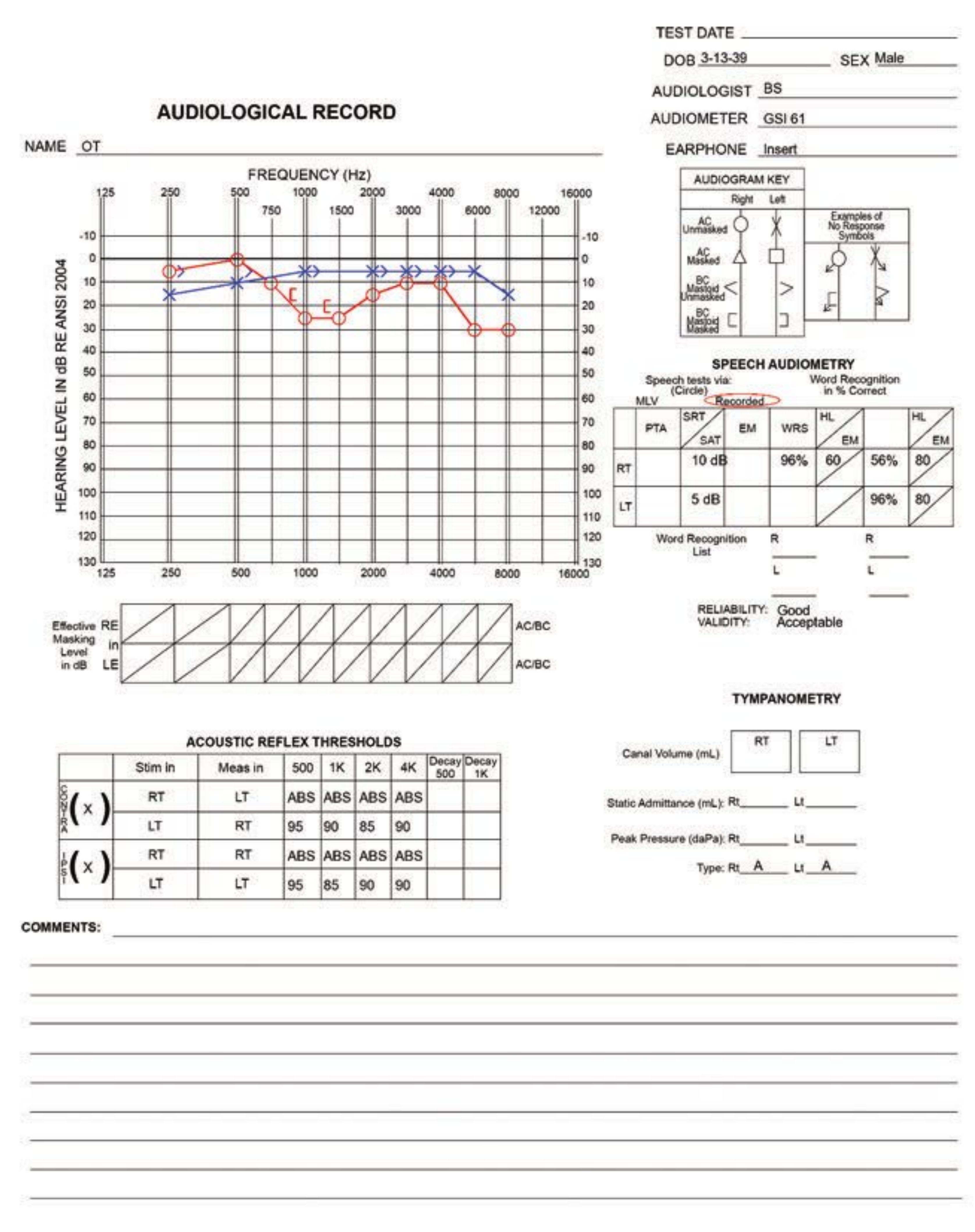
A 34 year-old male was reported as having mild hearing loss and tinnitus on his right ear.

13.1 Clinical History and Description

OT was 34 years old at the time of his initial clinical encounter. Three years prior to his audiological examination, he noticed right tinnitus and slight numbness over his right helix, following mild head trauma. These symptoms gradually subsided. Two weeks prior to his audiological examination, he noticed sudden mild hearing loss in his right ear. The hearing loss was accompanied by slight, constant tinnitus and numbness over his right auricle. The symptoms persisted, and he was referred for an audiological examination. OT denied dizziness, ear pain, fullness, or vegetative symptoms relating to the onset of the hearing loss and tinnitus.

13.2 Audiological Testing

Results of pure-tone audiometry are reported in Fig. 13.1. Left hearing sensitivity was within normal limits, with all thresholds being 15 dB HL or better across the audiometric frequency range. Right ear result showed a mild mid- and high-frequency sensorineural hearing loss. Word recognition scores (WRSs) were asymmetric as well. The WRS score for the left ear was 96% at 80 dB HL. For the right ear, the WRS was 96% at 60 dB HL, but decreased to 56% at 80 dB HL. This presence of "rollover" (i.e., decreased WRS as the presentation level is increased) of the word-recognition performance-intensity function is consistent with retrocochlear disorder on the right ear. Results from immittance audiometry were consistent with normal middle ear function bilaterally. The right ear had a normal, Type A tympanogram, normal static immittance, middle ear pressure and ear canal volume, and normal left crossed (i.e. contralateral) acoustic reflex thresholds. The left ear results were characterized by a normal,



AUDIOLOGICAL RECORD

NAME OT

TEST DATE

DOB 3-13-39 SEX Male

AUDIOLOGIST BS

AUDIOMETER GSI 61

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		10 dB		96%	60	56%	80
LT		5 dB				96%	80

Word Recognition List R L R L

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	95	90	85	90		
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	95	85	90	90		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt___ Lt___

Peak Pressure (daPa): Rt___ Lt___

Type: Rt A Lt A

COMMENTS:

Fig. 13.1 Pure-tone, speech audiometry, and immittance audiometry results in a 34-year-old man with symptoms of sudden, mild, right-sided hearing loss and tinnitus.

Type A tympanogram, normal static immittance, middle ear pressure and ear canal volume, and normal left uncrossed (i.e. ipsilateral) acoustic reflex thresholds. All acoustic reflex thresholds, however, with sound presented to the right ear (i.e. right crossed and right uncrossed) were absent. These results are consistent with a disorder of the right afferent portion of the reflex arc.

Transient-evoked otoacoustic emission (OAE) measures showed normal and symmetric emission amplitudes and reproducibility. The presence of symmetric OAEs in ears with asymmetric hearing is suggestive of retrocochlear disorder on the right side.

The overall pattern of results in the right ear is consistent with retrocochlear disorder, including asymmetric symptomatology, asymmetric hearing sensitivity, absent ARTs with contralateral and ipsilateral signals presented to the right ear, rollover of the word-recognition function, and preserved OAEs. The overall pattern of results in the left ear is consistent with normal function, including normal hearing sensitivity, ARTs, WRSs, and OAEs.

Following otologic examination, auditory brainstem response (ABR) testing was completed to assess auditory nervous system function. ABR results on the right ear were grossly abnormal. Only component Wave I could be recorded to alternating click stimuli presented at 80 dB nHL at 11.1 clicks per second. No replicable peaks could be identified beyond Wave I. ABR results on the left ear were also abnormal. Although all component waves were replicable, interwave latencies were abnormal. The Wave I to III interwave interval was 2.8 msec; the Wave III to V interwave interval was 2.0; the Wave I to V interwave interval was 4.8 msec, outside of three standard deviations from the mean. Middle and late latency responses were normal bilaterally.

13.3 Questions to the Reader

1. Is the presence of a slight asymmetry in hearing sensitivity by itself an indicator of the need to be concerned about retrocochlear pathology?
2. Does the sudden nature of the onset of the symptoms rule out an acoustic tumor or other eighth nerve disorder as a potential causative factor?
3. Rollover of word recognition seemed to be a useful screening measure in this case. How does a clinician quantify rollover?
4. Is there a more efficient method to evaluate the potential for rollover than measuring a complete performance-intensity function?
5. Results from the right ear reveal classic retrocochlear signs, including absent ARTs, OAEs present in the presence of hearing loss, rollover of the word-recognition performance function, and abnormal ABR. Results from the left ear do not. Are the abnormal left ear ABR results indicative of left-sided disorder or somehow related to the right-sided disorder?

13.4 Discussion of Questions to the Reader

1. Is the presence of a slight asymmetry in hearing sensitivity by itself an indicator of the need to be concerned about retrocochlear pathology?
   A tumor or other disorder of the acoustic nerve can cause asymmetric hearing loss. Asymmetric hearing loss per se, however, can be caused by any number of disorders, and most often the site of the disorder is the cochlea. Indeed, it may actually be the case that asymmetric hearing sensitivity loss is more normal than symmetric hearing sensitivity loss. Margolis and Saly, using a clinician-validated algorithm for defining asymmetry, reported that over half (55%) of patients with sensorineural hearing loss were classified as asymmetric. The authors concluded that asymmetric hearing loss has so many causes that no measure of asymmetry or formula for defining it will be sufficient to devise a screening test for acoustic tumors that will have acceptable sensitivity and specificity. Asymmetry in hearing sensitivity as a lone clinical sign is probably insufficient to arouse suspicion of a retrocochlear disorder. Corroborative audiometric indicators, however, can be powerful in determining the importance of an asymmetry. In the present case, the absence of ARTs and the presence of rollover of the word recognition function on the right ear accompanying asymmetric hearing thresholds provided ample evidence of auditory nervous system disorder.
2. Does the sudden nature of the onset of the symptoms rule out an acoustic tumor or other eighth nerve disorder as a potential causative factor?
   Most acoustic tumors are benign and grow slowly. Because of their slow growth, it is easy to imagine that the onset of symptoms would be gradual in nature. In fact, the majority of patients with this type of disorder do report a gradual onset of symptoms. A surprising percentage of patients, however, report that their initial symptoms occurred suddenly. Estimates suggest that between 15 and 19% of patients with an acoustic tumor have symptoms that appear suddenly. Thus nothing about the onset of symptoms in the present case would argue against the possibility of retrocochlear disorder.
3. Word recognition rollover seemed to be a useful screening measure in this case. How does a clinician quantify rollover?
   In normal ears and most ears with cochlear hearing loss, WRSs improve with increasing intensity until the word-recognition score asymptotes at a ceiling or maximum score. Increasing intensity does not change the WRS above that presentation level. In some ears with retrocochlear disorder, however, scores reach a maximum score as intensity is increased and then decrease at intensity levels above that maximum score. This so-called rollover can be very valuable as a screening tool when observed. The cause of rollover is unknown, but it is thought to be related to reduced nervous system temporal synchrony when the system is challenged at high intensity levels. Rollover has been associated with changes in nervous system function caused by discrete lesions of the eighth nerve and auditory brainstem and more diffuse deterioration, such as that relating to the aging process. Rollover is calculated as the difference in percentage between the lower-level maximum word-recognition score and the higher-level score, usually measured at 80 dB HL. Most clinicians use 20% as a rule of thumb for the presence of significant rollover. In most cases, rollover of less than 20% is considered to be consistent with cochlear disorder;

rollover of 20% or greater to be consistent with retrocochlear disorder. Another approach, intended to account for the absolute value of the maximum score, is to compute a rollover index (RI). This is calculated by dividing the amount of rollover by the maximum score. In the present case, the maximum score was 96%, and the minimum score was 56%. This results in a rollover index of 0.42 ([96 − 56]/96). The lower limit of for a normal RI varies among studies from 0.25 to 0.45. Regardless, in the case of OT, the presence of asymmetric rollover of 40% (i.e., 96% − 56%), in conjunction with the other retrocochlear signs, was sufficient to classify the rollover as abnormal.

4. Is there a more efficient method to evaluate the potential for rollover than establishing a complete performance-intensity function?
   Plotting a performance versus intensity function can be a time-consuming task, limiting its clinical utility. One way to approximate this approach is to screen for rollover. This is done by starting word-recognition testing at a high level of 80 dB HL. If the patient's WRS is greater than 80%, then significant rollover is ruled out. If the patient's WRS is poorer than 80%, the intensity can be reduced to determine if the WRS improves. This approach facilitates testing for rollover in a manner that is clinically efficient.
5. Results from the right ear reveal classic retrocochlear signs, including absent ARTs, OAEs present in the presence of hearing loss, rollover of the word-recognition performance function, and abnormal ABR. Results from the left ear do not. Are the abnormal left ear ABR results indicative of left-sided disorder or somehow related to the right-sided disorder?
   The abnormal ABR findings were not surprising given the clinical signs, and added more suspicion that the right disorder was retrocochlear in nature. The findings of abnormal ABR on the left ear were more surprising in that the behavioral and ART results were consistent with normal auditory nervous system function. Can an auditory nerve disorder, for example, an acoustic tumor, have such an extensive influence on brain function that it impacts the ABR from the contralateral ear? The answer is yes, but rarely. It appears that, on occasion, a tumor can influence the brainstem to such an extent that the function of the other side is affected and influences the ABR recording. When it happens, however, it usually affects the later waves of the ABR. In the case of OT, the disorder appears very early in the ABR as a prolongation of the Wave I to III interwave interval. These results suggest retrocochlear disorder on the left side as well as the right side, despite the difference in outcomes on the other measures.

13.5 Diagnosis and Recommended Treatment

Magnetic resonance imaging (MRI) was completed as a result of the audiometric and ABR findings consistent with retrocochlear disorder. One of the images is shown in Fig. 13.2. On the right side, a large 2 cm tumor was observed in the cerebellopontine angle (CPA), extending into the right internal auditory canal. Remarkably, an even larger tumor, 2.5 cm, was observed in the left CPA. In addition to these two tumors, two other large growths, an olfactory meningioma and a glomus jugularae tumor, were clearly identifiable on other MRI scans.

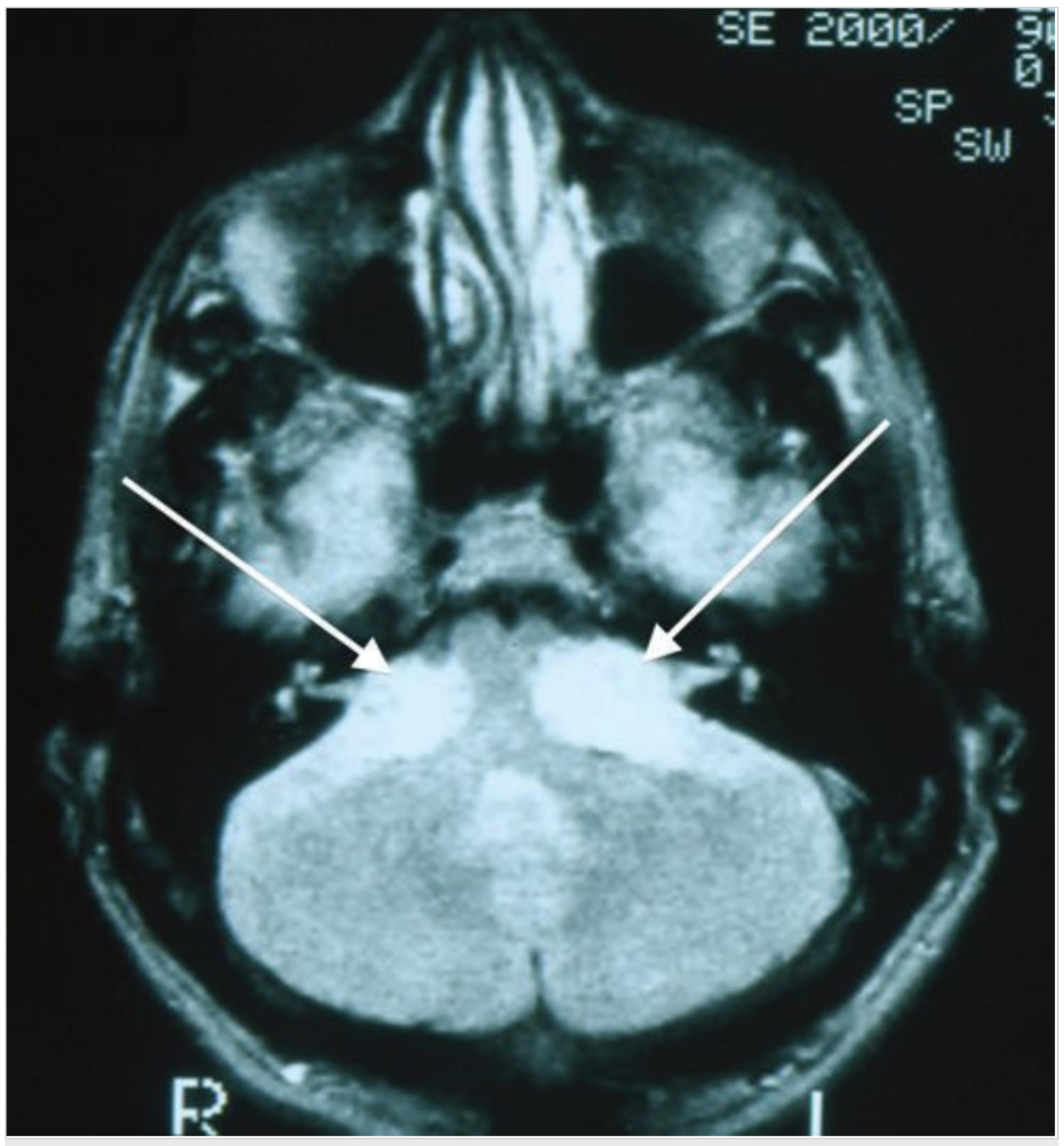
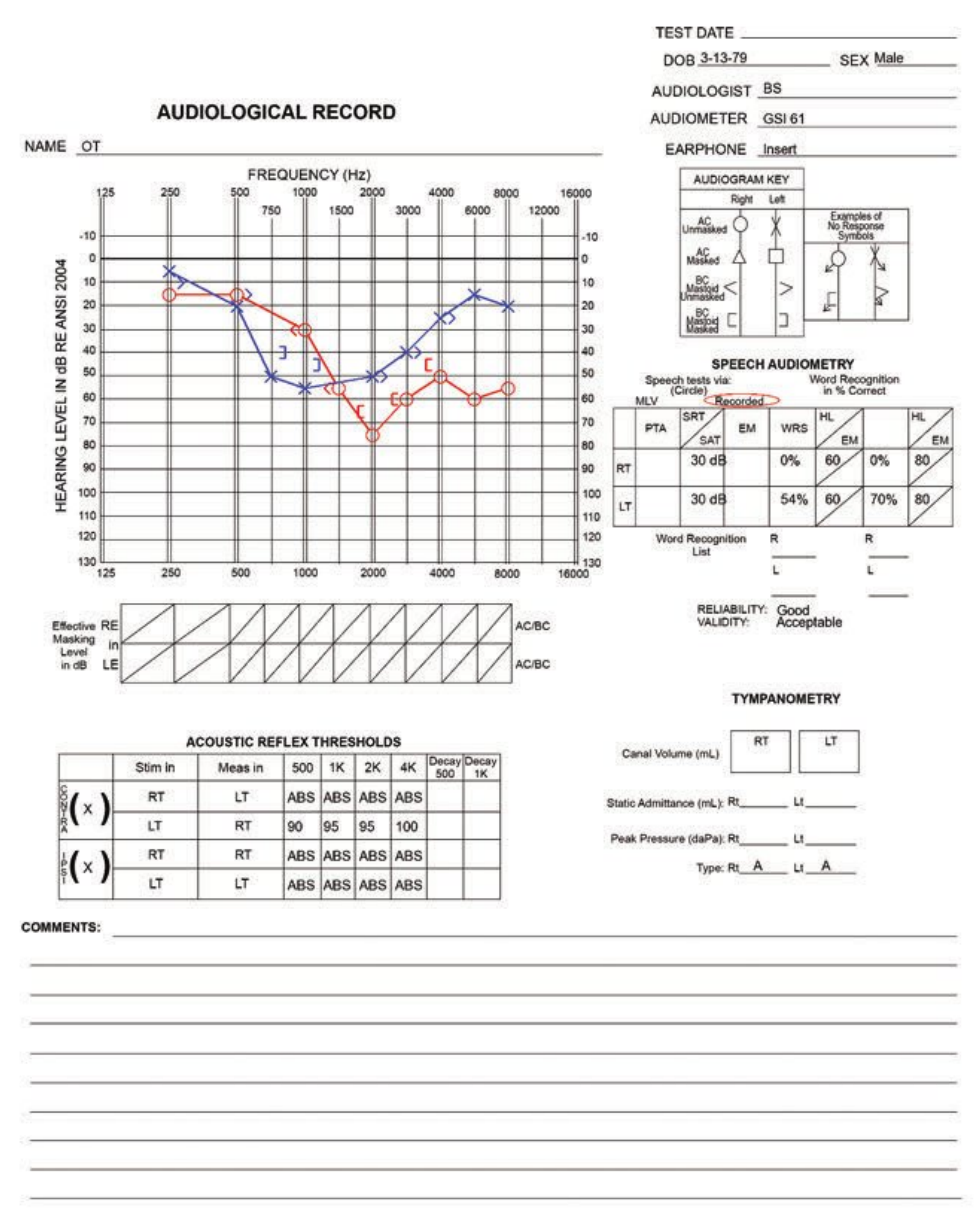


Fig. 13.2 Magnetic resonance imaging results in a 34-year-old man with bilateral acoustic tumors.

Based on results of the MRI, the patient was diagnosed with neurofibromatosis type 2 (NF2). NF2 is an inherited disorder, most commonly associated with bilateral vestibular schwannomas, although as in this case tumors can also form elsewhere in the brain. Because of the size of the tumors and the variety of locations, OT decided to forgo surgical treatment and let the disease process run its course.

13.6 Outcome

OT was last seen for follow-up audiological examination 5 years later at age 39 years. Results from the audiological examination are reported in Fig. 13.3. Left-ear hearing sensitivity decreased substantially since the initial examination five years earlier. There is now a moderate mid-frequency sensorineural hearing loss. Right ear results also showed a significant decrease, with a moderate to severe, high frequency sensorineural hearing loss. Word recognition scores continued to reveal substantial asymmetry. The maximum word recognition score for the left ear was 70% at 80 dB HL, which is consistent with the degree of hearing sensitivity loss. For the right ear, however, the word recognition score was 0% at all intensities tested. These results are poorer than would be predicted from the hearing loss and are reflective of the retrocochlear disorder. Immittance audiometry continued to show normal middle ear function on the right ear, characterized by a normal, Type A tympanogram, normal static immittance, middle ear pressure, and ear canal volume, and normal left crossed acoustic reflex thresholds. Acoustic reflex thresholds are plotted on the audiogram in Fig 1.3. The left ear results were characterized by a



AUDIOLOGICAL RECORD

NAME OT

TEST DATE

DOB 3-13-79 SEX Male

AUDIOLOGIST BS

AUDIOMETER GSI 61

EARPHONE Insert

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		30 dB		0%	60	0%	80
LT		30 dB		54%	60	70%	80

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt Lt
Peak Pressure (daPa): Rt Lt
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	90	95	95	100		
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS:

Fig. 13.3 Pure-tone, speech, and immittance audiometry results in the same patient at 39 years of age.

normal, Type A tympanogram and normal static immittance, middle ear pressure, and ear canal volume, although no acoustic reflex thresholds were recorded from the left probe. The absence of the left ipsilateral (uncrossed) ART is probably reflective of increased involvement of the left facial nerve.

The contrast of the effects of these tumors on the auditory outcomes remains remarkably asymmetric. To be sure, hearing sensitivity has decreased bilaterally, and the tumor is clearly now affecting the left ear. The effect, however, seems different in one important way. The right ear has all the clinical signs of a retrocochlear disorder, including absent ARTs and poorer than expected word recognition. The left ear, in contrast, has clinical signs that are more cochlear in nature, including the preservation of ART for left contralateral stimulation and WRSs more consistent with the degree of hearing sensitivity loss.

13.7 Additional Questions for the Reader

1. At the time of the initial evaluation, a tumor caused retrocochlear signs on one ear, but an even larger tumor did not seem to have a significant functional consequence on the opposite ear. How is that possible?
2. After 5 years, the overall pattern of results on the right ear was consistent with retrocochlear disorder, yet the overall pattern on the left ear seemed more consistent with cochlear disorder. How is that possible?

13.8 Discussion of Additional Questions for the Reader

1. At the time of the initial evaluation, a tumor caused retrocochlear signs on one ear, but an even larger tumor did not seem to have a significant functional consequence on the opposite ear. How is that possible?
   This case illustrates well the difference between structure and function relating to changes in the nervous system. OT has four sizable tumors in his brain, and the total impact is a mild hearing loss and tinnitus. These tumors are competing with his brain for space in four different regions, resulting in substantial structural changes, and yet the functional consequences are almost negligible. The opposite effect can also be

found in some cases. For example, in cases where an acoustic tumor is located entirely within the internal auditory canal, the tumor competes with the nerve for space in this narrow passageway. A very small tumor growing in this way can have a substantial primary effect on nerve function, resulting in significant functional auditory disorder. In this case, the tumor is growing out into the cerebellopontine angle, where it has more room to expand without obvious symptoms. With regard to OT's auditory system at the initial examination, the right acoustic tumor is impacting auditory function to a much more obvious extent than the left acoustic tumor. The right acoustic tumor is causing auditory symptoms, a hearing sensitivity loss, elimination of ARTs, impaired WRSs, and disordered synchronous neural activity of the eighth nerve. The left acoustic tumor does not have the same effect. There are no auditory symptoms, no measurable hearing sensitivity loss, no loss of contralateral ARTs, and no deficits in WRS. There is some evidence of disorder, manifesting in prolonged ABR Wave I to III interwave intervals and absent left ipsilateral, but even this does not seem to reflect changes in perceived hearing ability. This case serves to illustrate, in a single patient, the differences that can be observed in the functional consequences of structural changes.

2. After five years, the overall pattern of results on the right ear was consistent with retrocochlear disorder, yet the overall pattern on the left ear seemed more consistent with cochlear disorder. How is that possible?
   When a tumor has a primary influence on function of the eighth nerve, retrocochlear signs are expected. These may include hearing sensitivity loss, poorer than expected WRSs, auditory adaptation, absent or elevated ARTs, abnormal ABR, and so on. Sometimes, however, clinical signs are observed that are more consistent with cochlear disorder. The conventional wisdom for the cause of such outcomes is that the tumor is somehow interfering with the cochlea's blood supply, resulting in changes in cochlear function. That may not be the case. Mahmud et al provided some excellent insight into an alternative cause of cochlear disorder in some of these patients. They evaluated the temporal bones of 11 cases of unoperated unilateral acoustic tumors. They quantified tumor size, spiral ganglion cells, hair cells, stria vascularis, and the spiral ligament. Not unsurprisingly, they found that increased tumor size predicted a reduced spiral ganglion count. They also found, however, that in some cases, there was significant degeneration of cochlear structures, including hair cells, stria vascularis, and spiral ligament. They concluded that acoustic tumors can cause hearing loss both by degeneration of the auditory nerve and by inducing degenerative changes to the inner ear. In OT, these two influences may be affecting the two ears differentially. On the right ear, the tumor appears to be exerting a primary effect on acoustic nerve function, resulting in retrocochlear signs. On the left ear, over time, the tumor appears to be exerting a secondary effect on cochlear function, resulting in cochlear signs.

13.9 Key Points

1. The influence of structural changes in the auditory nervous system can result in varied functional outcomes.
2. The authors present a case in which bilateral tumors affect hearing ability distinctly between the two ears.
3. Clinical outcomes in this patient appear to show a primary effect of an acoustic tumor on nerve function in one ear and a secondary influence of an acoustic tumor on cochlear function in the other ear.

Suggested Reading

Mahmud MR, Khan AM, Nadol JB. Histopathology of the inner ear in unoperated acoustic neuroma. Ann Otol Rhinol Laryngol 2003; 112: 979–986

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Pensak ML, Glasscock ME, Josey AF, Jackson CG, Gulya AJ. Sudden hearing loss and cerebellopontine angle tumors. Laryngoscope 1985; 95: 1188–1193

Yanagihara N, Asai M. Sudden hearing loss induced by acoustic neuroma: significance of small tumors. Laryngoscope 1993; 103: 308–311

14 Musical Hallucinations

Jennifer Stockwell and Ross J. Roeser

A patient is referred because he reports hearing music in his right ear continuously, following a transient ischemic attack.

14.1 Clinical History and Description

RF is a 72-year-old male who leads an extremely active lifestyle. He jogs regularly, bikes, swims or exercises daily, and is in very good general physical health. He has, however, a significant medical history that includes hyperlipidemia, high blood pressure, and tachycardia, for which he takes daily medications.

While exercising one morning with friends, RF experienced temporary weakness, numbness, and loss of motor control of the right side of his body. He did not seek immediate medical attention but went to the emergency room the following afternoon. The patient was counseled that he likely experienced a transient ischemic attack (TIA), though an official diagnosis was never made due to lack of physiological evidence.

Approximately 2 to 3 days following the TIA, RF began hearing constant music, seemingly originating from his right ear, that has been continuous. The music typically follows the same patterns and rarely has any fluctuations. Characteristics of the music include a male chorus, songs that are familiar to him from his childhood ("Take Me Out to the Ball Game," "Silent Night," "Easter Parade," etc.), and is of constant intensity, though less noticeable in more background noise. RF has the capability to change the song by thinking of another song and indicated the music often sounds like a broken record as if the verse is unable to finish and constantly starts over. He also complains of tinnitus in the left ear. The tinnitus and musical perceptions typically occur simultaneously.

RF underwent neurological and audiological examinations in an effort to identify the origin of the perceived music and find ways to manage the condition. The neurological examination included magnetic resonance imaging (MRI), electrocardiogram (EKG), electroencephalogram (EEG), and magnetic resonance angiography (MRA). Results for these examinations were reported to be within normal limits or unremarkable with respect to his current symptoms.

14.2 Audiological Testing

As shown in Fig. 14.1, pure-tone thresholds revealed a bilateral moderate reversed trough-shaped (i.e., better hearing at 1000 to 4000 Hz than the frequency region below and above) sensorineural hearing loss with the right ear having slightly better low frequency hearing thresholds than the left ear. Speech recognition thresholds (SRT) were within normal limits bilaterally and in good agreement with pure-tone average (PTA). Word recognition scores (WRSs), obtained at 85 dB HL revealed slight difficulty in the ability to recognize speech in the right ear and normal ability to recognize speech in the left ear. Otoscopy was unremarkable. Tympanometry revealed normal static admittance (mL), middle ear pressure (daPa), and ear canal volume (mL) bilaterally. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present at expected hearing levels bilaterally. Speech-in-noise testing using the Quick SIN at 70 dB HL yielded a 3.5 dB signal to noise ratio (SNR) loss for the left ear and a 3.0 dB SNR loss for the right ear, indicating a mild SNR loss. Residual inhibition was assessed by presenting speech noise binaurally via earphones at 80 dB HL for 60 seconds. This intensity level, or higher levels, failed to mask RF's musical perceptions either during the presentation of the masking signal or after masking was discontinued. These observations indicate that no residual inhabitation could be demonstrated.

As shown in Fig. 14.2, distortion-product otoacoustic emissions (DPOAEs) were present and robust at 750 to 8000 Hz bilaterally. Fig. 14.3 shows that spontaneous OAEs (SOAEs) were absent in the left ear and were present in the right ear. This finding is somewhat surprising and of interest in that RF reported hearing music from his right ear. Of note is that RF indicated he perceived the music during the SOAE test.

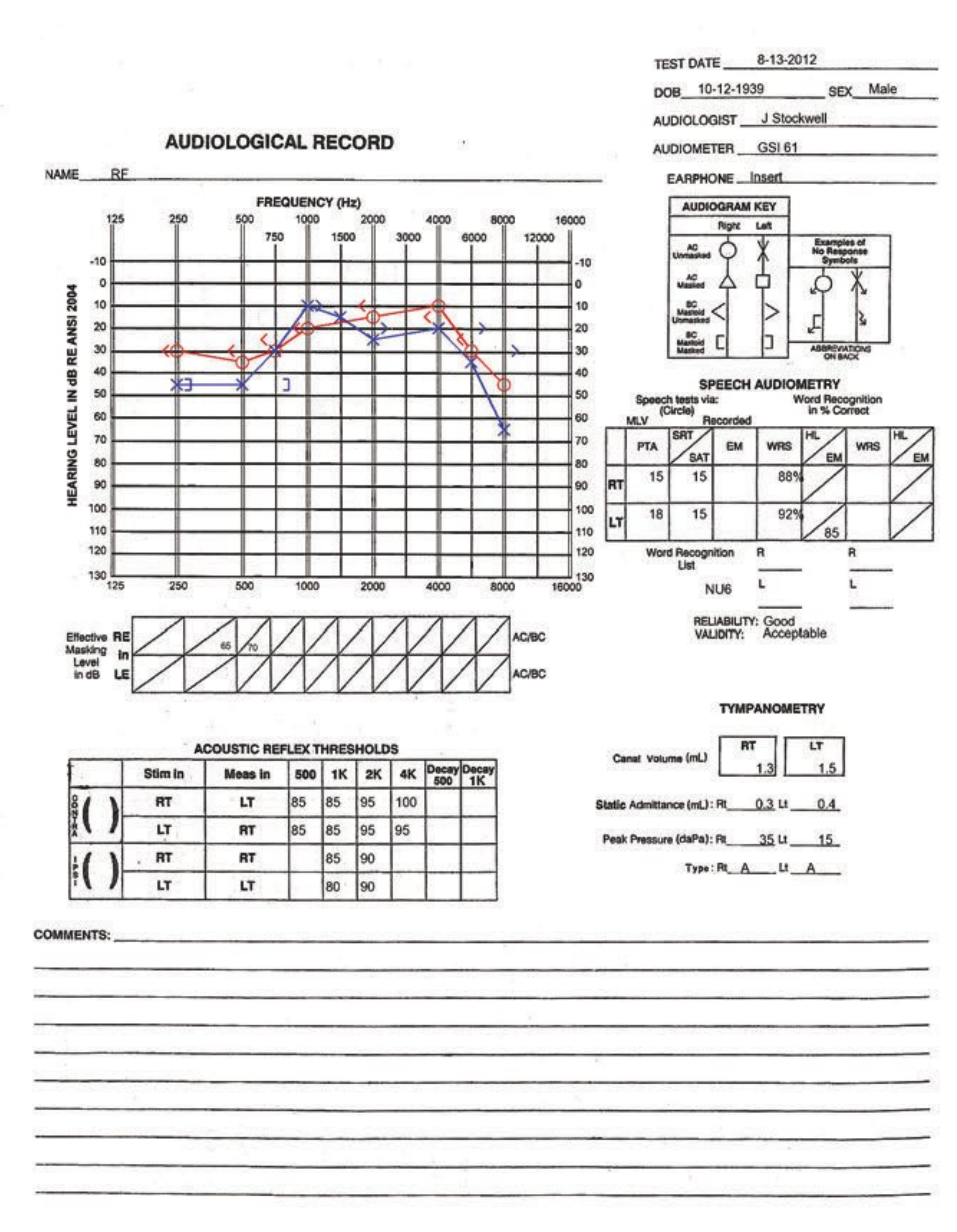
The patient was a pilot participant in a research study in which vestibular testing was performed on all subjects. Videonystagmography (VNG) included ocular motor procedures including gaze, saccades, pursuit, and optikinetic testing; all of the test results were within normal limits. Positioning/positional testing included the Dix-Hallpike maneuver, sitting, supine, head right, head left, body right, and body left. No nystagmus was identified in any position. Bithermal caloric testing was also within normal limits, yielding neither a unilateral weakness nor a directional preponderance.

14.3 Questions to the Reader

1. How are this patient's symptoms different from his having "a song stuck in his head"?
2. Does the presence of SOAEs have significance for patients who are experiencing auditory or musical hallucinations?
3. Do musical or auditory hallucinations stem from a peripheral or a central site of lesion?
4. What is the role of audiology in the diagnosis and treatment of auditory/musical hallucinations?
5. What possible medical and audiological treatment/management options are there for patients with auditory hallucinations?

14.4 Discussion of Questions to the Reader

1. How are this patient's symptoms different from his having "a song stuck in his head"?
   Many individuals after hearing a song report that they keep thinking of the song, or the song becomes "stuck in their head." This condition has been referred to as "having an earworm." Other terms used to describe this perception include musical imagery repetition and involuntary musical imagery.


FREQUENCY (Hz)
125 250 500 1000 2000 4000 8000 16000
750 1500 3000 6000 12000
HEARING LEVEL IN dB RE ANSI 2004
-10 0 10 20 30 40 50 60 70 80 90 100 110 120 130
AUDIOGRAM KEY
Right Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

AUDIOLOGICAL RECORD

NAME RF

TEST DATE 8-13-2012
DOB 10-12-1939 SEX Male
AUDIOLOGIST J Stockwell
AUDIOMETER GSI 61
EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	15	15		88%			
LT	18	15		92%	85		

Word Recognition List R ____ L ____ R ____ L ____
NU6

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB											
RE in		65	70								AC/BC
LE											AC/BC

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.3	1.5

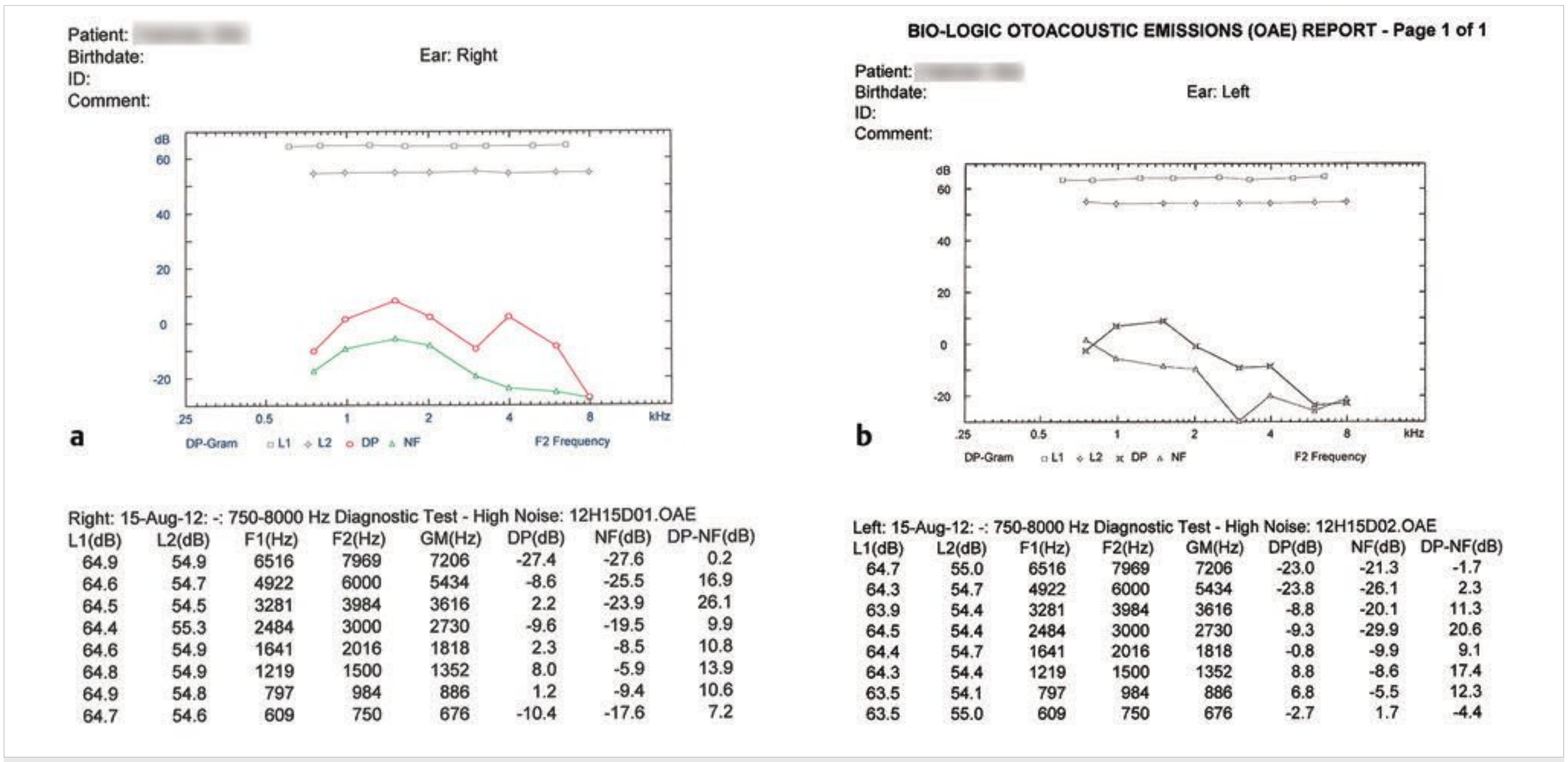
Static Admittance (mL): Rt 0.3 Lt 0.4
Peak Pressure (daPa): Rt 35 Lt 15
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	85	85	95	100		
	LT	RT	85	85	95	95		
IPSI ( )	RT	RT		85	90			
	LT	LT		80	90			

COMMENTS:

Fig. 14.1 Audiological examination for RF experiencing musical hallucinations following a left hemisphere transient ischemic attack (TIA).



Patient: [redacted]
Birthdate:
ID:
Comment:
Ear: Right

```
Right: 15-Aug-12: -: 750-8000 Hz Diagnostic Test - High Noise: 12H15D01.OAE
L1(dB)   L2(dB)   F1(Hz)   F2(Hz)   GM(Hz)   DP(dB)   NF(dB)   DP-NF(dB)
 64.9     54.9     6516     7969     7206    -27.4    -27.6        0.2
 64.6     54.7     4922     6000     5434     -8.6    -25.5       16.9
 64.5     54.5     3281     3984     3616      2.2    -23.9       26.1
 64.4     55.3     2484     3000     2730     -9.6    -19.5        9.9
 64.6     54.9     1641     2016     1818      2.3     -8.5       10.8
 64.8     54.9     1219     1500     1352      8.0     -5.9       13.9
 64.9     54.8      797      984      886      1.2     -9.4       10.6
 64.7     54.6      609      750      676    -10.4    -17.6        7.2
```

BIO-LOGIC OTOACOUSTIC EMISSIONS (OAE) REPORT - Page 1 of 1

Patient: [redacted]
Birthdate:
ID:
Comment:
Ear: Left

```
Left: 15-Aug-12: -: 750-8000 Hz Diagnostic Test - High Noise: 12H15D02.OAE
L1(dB)   L2(dB)   F1(Hz)   F2(Hz)   GM(Hz)   DP(dB)   NF(dB)   DP-NF(dB)
 64.7     55.0     6516     7969     7206    -23.0    -21.3       -1.7
 64.3     54.7     4922     6000     5434    -23.8    -26.1        2.3
 63.9     54.4     3281     3984     3616     -8.8    -20.1       11.3
 64.5     54.4     2484     3000     2730     -9.3    -29.9       20.6
 64.4     54.7     1641     2016     1818     -0.8     -9.9        9.1
 64.3     54.4     1219     1500     1352      8.8     -8.6       17.4
 63.5     54.1      797      984      886      6.8     -5.5       12.3
 63.5     55.0      609      750      676     -2.7      1.7       -4.4
```

Fig. 14.2 Distortion-product otoacoustic emissions for RF's right ear and left ear.

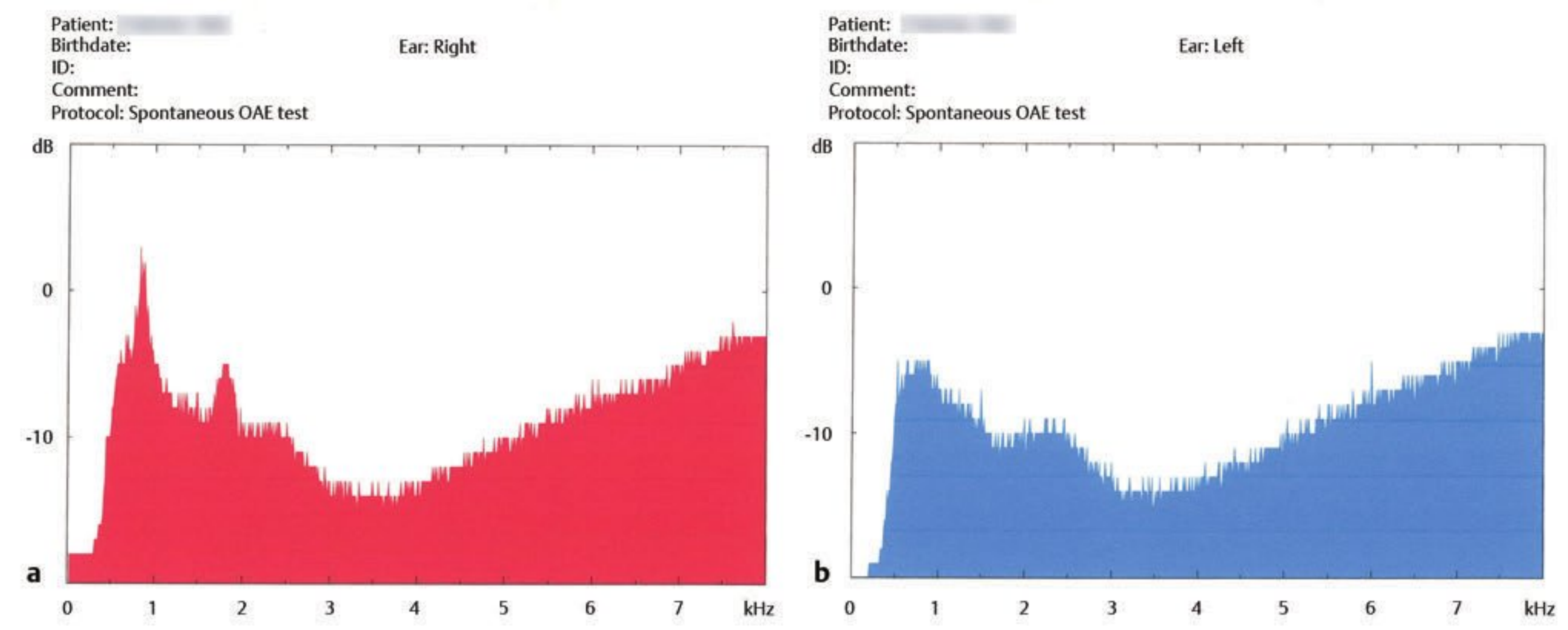


Fig. 14.3 Spontaneous otoacoustic emissions for RF's right ear and left ears.

Earworms, however, do not have the saliency or reality of a true perception; the experience is common in everyday life and is distinguished from brain damage, which results in the abnormal form of musical perception, referred to as palinacousis. Palinacousis is an auditory form of perseveration that is described as one's continuing to hear a sound after the physical noise has disappeared. This perception is often associated with lesions of the temporal lobe.

A phenomenon that is more severe than an earworm and, can cause the patient to believe the perceived sound is real is auditory hallucination. Generally, a hallucination is the experience of a perception without a corresponding external stimulus. The prevalence of auditory hallucination is common (10–30 cases per 1000 people per year when all sensory systems are included). Auditory hallucinations are primarily referred to as tinnitus and may be described as buzzing, ringing, sizzling, or other sensations. They are a common subtype of hallucinations.

Musical hallucinations are a type of auditory hallucination, described as subjective experience of hearing music. Experiencing musical hallucinations stems from a disorder of processing complex sounds. These hallucinations are typically short fragments of simple melodies, often from music heard regularly, such as hymns and carols, and familiar from youth. Musical hallucinations are phenomenologically distinct from verbal hallucinations and have different neurological correlates. Specifically, musical hallucinations are significantly less common than auditory hallucinations and represent a small portion of hallucinations (0.16% of admitted psychiatric patients). Musical hallucinations are more common (70–80%) in women over 60 years of age who are socially isolated. In summary, an earworm can be described as having a song "stuck" in one's head. This nuisance is short lived and a common phenomenon. Musical hallucinations, however, are a real perception of hearing music when no music is present. Musical hallucinations are rare, long-lasting, and have the potential to adversely affect quality of life.

2. Does the presence of SOAEs have significance for patients who are experiencing auditory or musical hallucinations?
The presence of SOAEs in the right ear, where RF perceives music, but not in the left ear, was an unexpected finding. RF was counseled on the physiology of the cochlea and how an "echo" is produced as a result of stimulating the ear. Likewise, in the absence of a stimulus, no echo should be produced because the cochlea has not been stimulated. The authors of this case report were expecting to measure no response for either ear and use this finding as a counseling tool to describe the hallucination to be stemming from the cortex and not from the periphery. A robust response was observed between 1000 and 2000 Hz for the right ear. RF did indicate he was hearing music during the test. As expected, no response was observed for the left ear. This test was repeated 2 weeks later and yielded the same result.

SOAEs occur in approximately 50% of the population, primarily women, and originate from the outer hair cells corresponding to the test frequency. More research is needed to further investigate the pathophysiology of these emissions as well as to identify their clinical utility. There is extensive research to support a link between SOAEs and tinnitus; however, there are also many studies that refute this evidence. These contradictory findings encourage the need for more research on SOAEs.

It is well accepted that SOAEs will not be present where sensorineural hearing loss exceeds 30 dB HL. The presence of SOAEs in RF's right ear and not in the left ear may be the result of better hearing in his right ear.

In summary, the result of SOAEs present in the right ear that perceives music and absent in the left ear is a curious finding. Though the SOAEs were likely recorded due to better hearing in this right ear, more research regarding SOAEs in the presence of abnormal auditory perceptions needs to be conducted.

3. Do musical hallucinations originate from a peripheral or a central site of lesion?
Evidence exists to support both peripheral and central sites of lesion, although it is more likely that the hallucinations originate from a central pathology. There is a possible relationship between musical hallucinations and hearing loss,

but research is inconclusive. Many cases of musical hallucinations have been reported in patients with severe or profound hearing loss, suggesting peripheral pathology is a likely cause of these hallucinations. There is, however, limited research regarding mild hearing loss and the presence of these hallucinations. Similarly, cases identifying musical hallucinations also report confounding psychological disturbances that may be influencing the perception.

Evidence to suggest the hallucinations originate from a central pathology is rather extensive. There are multiple reports that identify damage to critical brain regions that leads to musical hallucinations. Such brain regions include the brainstem, pons, thalamus, and auditory radiations. Many patients who experience musical hallucinations also report a psychological disorder. Depression is present in 40% of these patients, as well as obsessive compulsive disorder, schizophrenia, and neurotic symptoms.

Research has been reported to support peripheral, central, and psychological abnormalities as the primary cause of musical hallucinations. Because RF began experiencing his hallucinations following a suspected TIA, the most likely cause of these hallucinations is a central site of lesion. Peripheral and psychological factors may contribute to the perception, but it is likely reorganization and plasticity of cortical structures following the TIA that triggered RF’s musical hallucinations.

4. What is the role of audiology in the diagnosis and treatment of auditory/musical hallucinations?
   Because the etiology of musical hallucinations is questionable and seems to vary from patient to patient, it is necessary to have appropriate referral sources for patients who may present with these symptoms. Involvement of neurologists, otologists, and psychologists is important for these patients. Diagnosis and treatment of musical hallucinations will likely result from input from each of these disciplines, emphasizing the necessity for a team approach. Because musical hallucinations often occur with hearing loss, audiological management will often be an advantageous treatment option. The amelioration of hearing loss with amplification is an appropriate first-line treatment. Likewise, sound generators to mask the musical perception or combination units (i.e., devices that are hearing aids and maskers) to treat hearing loss and provide masking may also be effective.
5. What possible medical and audiological treatment/management options are there for patients with auditory hallucinations?
   Medical treatment may include drugs to reduce the perception of hallucinations. Anticonvulsant and antidepressive agents have been effective in the treatment of some musical hallucinations. Such drugs have proven effective in reducing the effects of coexisting psychiatric disorders and reducing the effect of musical hallucinations. Audiological treatment options include hearing aids (when hearing loss is present), sound generators, tinnitus treatment devices, and auditory rehabilitation. It is necessary to identify the severity of the perceptions and the patient’s primary needs and develop treatment options accordingly based on the individual’s primary concerns.

14.5 Diagnosis and Recommended Treatment

RF was diagnosed with a bilateral asymmetrical (right ear better than left ear) mild to moderate trough-shaped sensorineural hearing loss. Despite having only a mild loss of hearing sensitivity, RF reported significant difficulty recognizing speech in quiet and noise. Based on his subjective difficulty with speech recognition, as well as his musical hallucinations, he was fit bilaterally with mild gain digitally programmable behind-the-ear (BTE) thin-tube hearing instruments. After wearing the hearing aids for 3 months RF reported significant improvement in his ability to recognize speech, but no or little reduction or diminution of his musical hallucinations. The use of masking generators was considered, but because external stimuli from sound sources had little or no effect on the musical perceptions and/or residual inhibition could not be demonstrated, masking generators were excluded from the fitting.

14.6 Outcome

RF elected to continue wearing his hearing instruments. He was given the option of neurological follow-up with pharmacological treatment but declined due to the possible side effects of the medications. He stated he gradually has slowly habituated to his musical perceptions and felt that he was able to “live with them.” Compared with his initial perception, he rated his present perception as being about half as annoying.

The presence of SOAEs in the right ear, where RF perceives music, was unexpected. Because this is the first case to report this finding, other such reports are needed to investigate if there is a relationship between SOAEs and musical hallucinations.

14.7 Key Points

1. Musical hallucinations are rare. When they exist, audiologists should be involved in the care of the patient to define the degree and type of hearing loss as well as provide a first line of treatment if hearing loss is found.
2. Intervention with hearing aids should be provided for patients with musical hallucinations who present with hearing loss.
3. The finding of SOAEs in the ear with musical hallucinations was unexpected and should be investigated in future patients.

Suggested Reading

Cole MG, Dowson L, Dendukuri N, Belzile E. The prevalence and phenomenology of auditory hallucinations among elderly subjects attending an audiology clinic. Int J Geriatr Psychiatry 2002; 17: 444–452

Gordon AG. Do musical hallucinations always arise from the inner ear? Med Hypotheses 1997; 49: 111–122

Jacobs L, Feldman M, Diamond SP, Bender MB. Palinacousis: persistent or recurring auditory sensations Cortex 1973; 9: 275–287

Tanriverdi N, Sayilgan MA, Ozçürümez G. Musical hallucinations associated with abruptly developed bilateral loss of hearing. Acta Psychiatr Scand 2001; 103: 153–155

Williamson VJ, Jilka SR, Fry J, Finkel S, Müllensiefen D, Stewart L. How do “earworms” start? Classifying the everyday circumstances of involuntary musical imagery. Psychol Music 2012; 40: 259–284

15 Multiple Pathologies in a Single Patient

Sarah A. Sydlowski

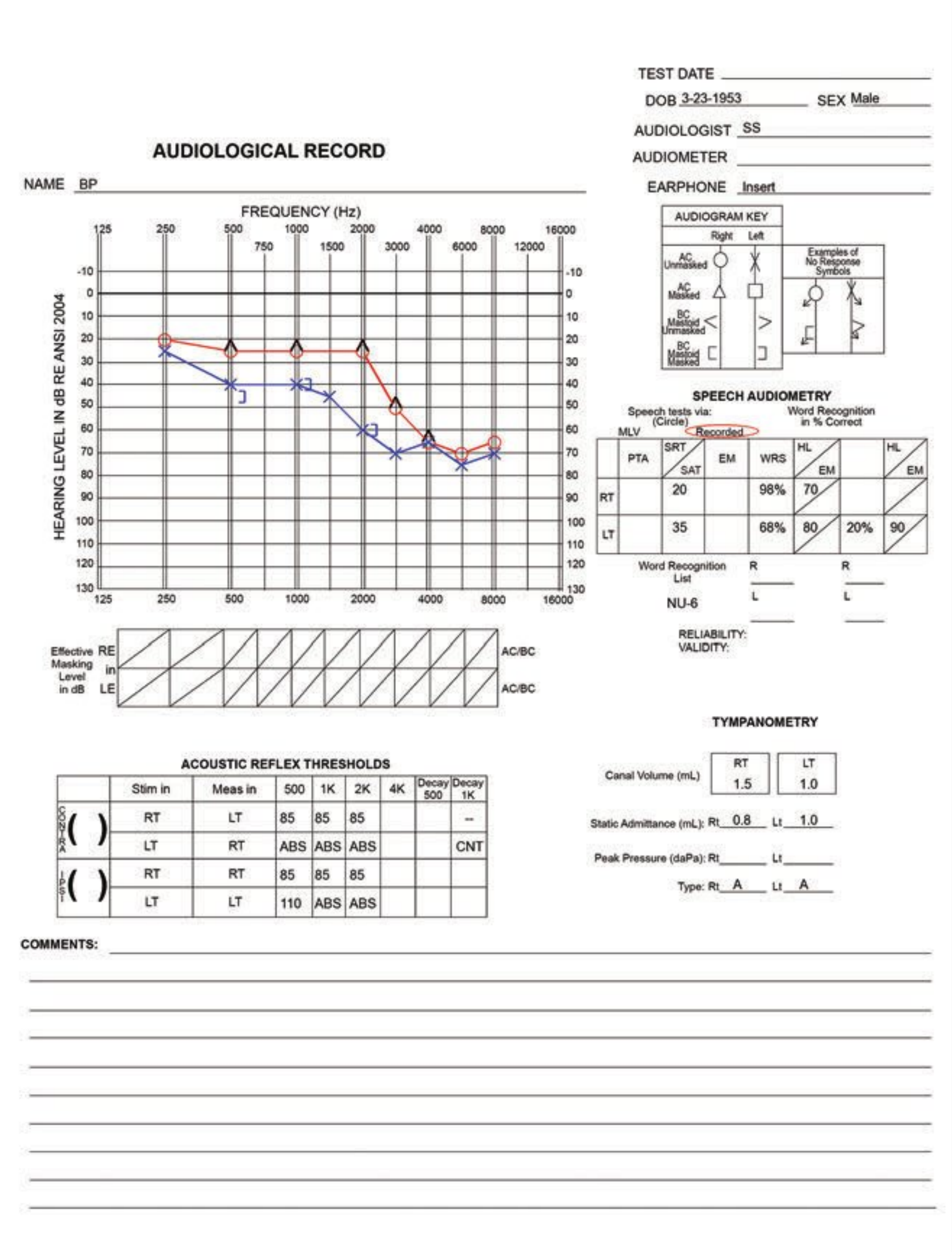
A 60-year-old male presented with a complex medical history. His current case history included left-sided “sizzling” tinnitus and perceived decrease in hearing following treatment with cisplatin for mesothelioma.

15.1 Clinical History and Description

BP was referred for audiological examination due to constant, “sizzling” tinnitus in his left ear for the past 3 months that began concurrently with the initiation of cisplatin chemotherapy for the treatment of mesothelioma of the left upper thorax. BP also reported decreased hearing bilaterally. The patient had a positive history of occupational noise exposure without the use of hearing protection. He reported that he has, however, used hearing protection consistently since beginning his current course of chemotherapy.

15.2 Audiological Testing

The results from the initial audiological examination (□ Fig. 15.1) revealed an asymmetric sensorineural hearing loss (SNHL). Results for the right ear revealed a slight SNHL at 250 to 2000 Hz sloping to a moderate to moderately severe SNHL at 3000 to 8000 Hz. Results for the left ear, on the other hand, revealed a mild SNHL at 500 to 1500 Hz decreasing to a moderately severe SNHL at 2000 to 3000 Hz. Thus there was a significant asymmetry in hearing between the right and left ears at 500 to 3000 Hz, with the left ear being poorer than the right ear. In addition, the speech recognition threshold (SRT) for the right ear revealed a slight loss in the ability to receive speech, whereas results for the left ear revealed a mild loss in the ability to receive speech. The word recognition score (WRS) in the right ear was 98%, whereas the WRS for the left ear was 68% and decreasing to 20% when the presentation level was increased by 10 dB HL. Bilateral tympanograms revealed normal



AUDIOLOGICAL RECORD

NAME BP

TEST DATE

DOB 3-23-1953 SEX Male

AUDIOLOGIST SS

AUDIOMETER

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT		20		98%	70		
LT		35		68%	80	20%	90

Word Recognition List NU-6 R L R L

RELIABILITY:
VALIDITY:

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.5	1.0

Static Admittance (mL): Rt 0.8 Lt 1.0

Peak Pressure (daPa): Rt Lt

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	85	85	85			--
	LT	RT	ABS	ABS	ABS			CNT
IPSI ( )	RT	RT	85	85	85			
	LT	LT	110	ABS	ABS			

COMMENTS:

Fig. 15.1 Results from the current audiological examination.

ear canal volume (mL), middle ear pressure (daPa), and peak compliance (mL). Ipsilateral acoustic reflex thresholds (ARTs) to the right ear were normal, whereas ipsilateral ARTs to the left ear were elevated at 500 Hz and absent at 1000 and 2000 Hz. Contralateral SRTs to the right ear were normal, whereas contralateral ARTs to the left ear were absent at 500 to 2000 Hz.

15.3 Questions to the Reader

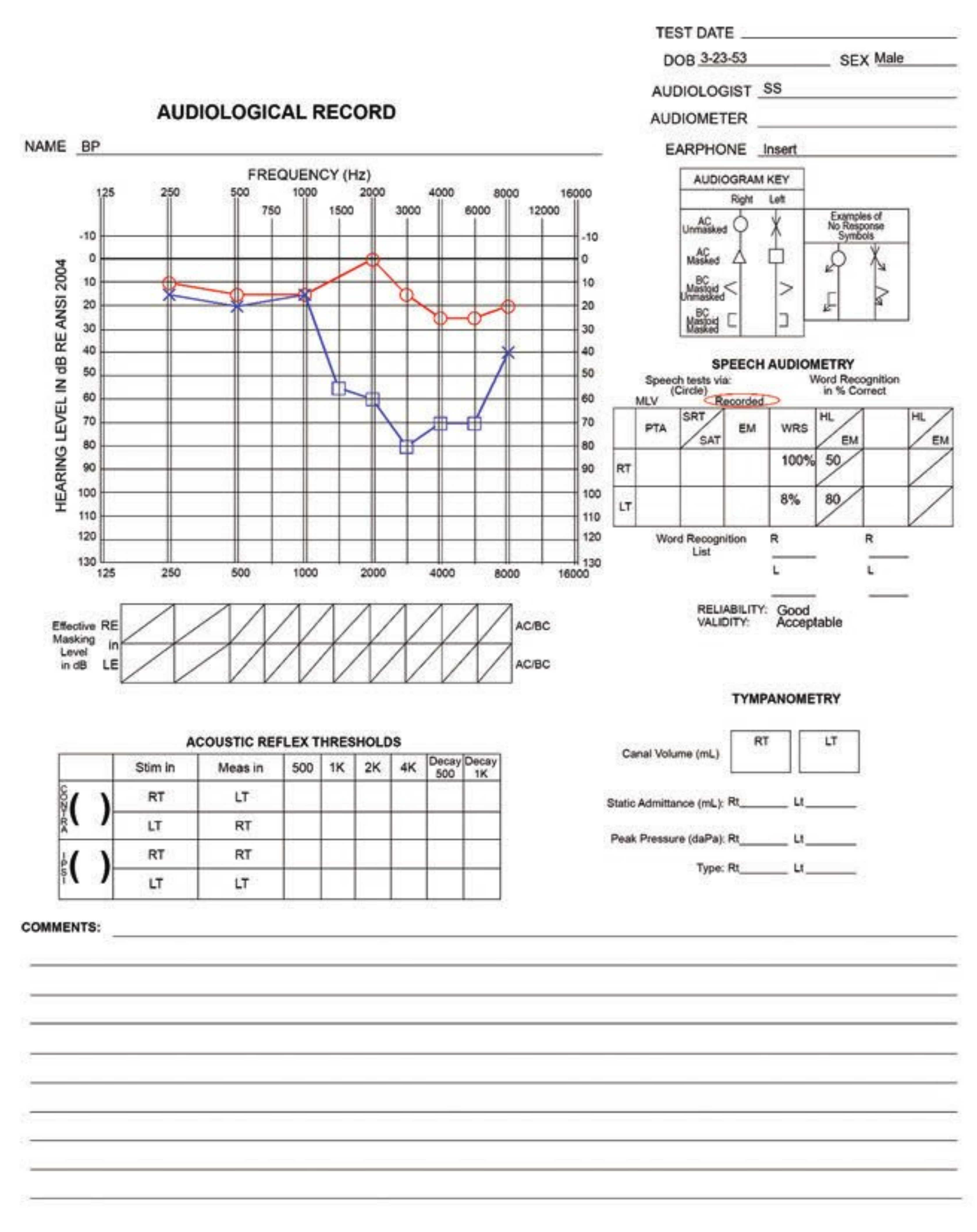
1. Are the results from the audiological examination consistent with what a clinician might expect given the history of administration of an ototoxic chemotherapeutic agent?
2. Describe the ARTs. Are the ARTs consistent with the audiometric results?
3. Consider the results of speech audiometry. Which factors, if any, in the patient's history might account for these results?
4. Given the results of the audiological examination, what further information is required?

15.4 Discussion of Questions to the Reader

1. Are the results from the audiological examination consistent with what a clinician might expect given the history of administration of an ototoxic chemotherapeutic agent?
   Cisplatin is known to be extremely and predictably ototoxic. Susceptibility to ototoxicity increases with certain risk factors that may include age, preexisting hearing loss, and prior cranial irradiation. Patients with cisplatin ototoxicity typically present with bilateral symmetrical high-frequency SNHL and tinnitus. Hearing loss resulting from cisplatin ototoxicity may progress to the lower frequencies and could be asymmetrical. Notably, WRSs may be significantly reduced. Given this information, a bilateral symmetrical SNHL would have been predicted in this case. Significant asymmetry, however, as reported in Fig. 15.1, was noted.
2. Describe the ARTs. Are the ARTs consistent with the audiometric results?
   The site of lesion in cisplatin ototoxicity is believed to be the hair cells and stria vascularis of the cochlea. When administered in combination with radiation, spiral ganglion cells may also be involved. In cases of cochlear hearing loss, ARTs are typically present at reduced sensation levels until the hearing loss exceeds approximately 70 dB HL, at which point ARTs are typically absent. In this case report, BP's ARTs are elevated or absent when stimulation is presented to the left ear. If the hearing loss were cochlear as would be expected due to cisplatin ototoxicity, the ARTs should have been present and at reduced sensation levels.
3. Consider the results of speech audiometry. Which factors, if any, in the patient's history might account for these results?
   A maximum WRS (PB_{max}) is typically measured at approximately 30 to 40 dB sensation level (SL) regarding SRT or at the patient's most comfortable listening level (MCL). In cochlear hearing loss, the WRS is generally reduced in a predictable pattern consistent with the magnitude and configuration of the hearing loss. As the presentation level increases, WRS typically improves. In cisplatin ototoxicity, which has a primarily cochlear site of lesion, clinicians would anticipate the WRS to be consistent with the magnitude and configuration of the hearing loss. Due to the asymmetric nature of BP's audiogram, a clinician might anticipate asymmetric WRSs. Indeed this expectation is observed (i.e., 96%for the right ear compared with 68%for the left ear). Additionally, however, BP's WRS decreased with increased presentation level (i.e., 68%to 20%when the presentation level was increased by 10 dB HL), and this “rollover” is inconsistent with cochlear hearing loss and cisplatin ototoxicity. Rollover has been reported to differentiate cochlear from retrocochlear lesions by calculating the rollover index (RI). The RI is calculated using a performance-intensity (PI) function (i.e., measure WRS at increasing presentation levels) obtained with phonetically balanced words. The best word recognition score (PB_{max}) and the poorest WRS (PB_{min}) are then compared using the calculation $(PB_{max} - PB_{min})/PB_{max}$. Significant RI values for differentiation between cochlear and retrocochlear disorders have ranged from ≥ 0.25 to ≥ 0.45. In BP's case, the RI is 0.71, which is clinically significant and indicative of retrocochlear involvement.
4. Given the results of the audiological examination, what further information is required?
   The results of the initial audiological examination include several inconsistencies that are incongruous with cisplatin ototoxicity. Specifically, although cisplatin ototoxicity may cause decreased WRS and asymmetric hearing loss, BP also exhibits further decreases in WRS at higher presentation levels (i.e., RI) as well as a “stimulus effect” ART pattern that is consistent with retrocochlear involvement of the left ear. Further information that may be relevant to BP's case includes obtaining a more comprehensive case history. This information should include hearing loss thresholds prior to the administration of cisplatin, magnetic resonance imaging (MRI), or the results of an auditory brainstem response (ABR) test.

15.5 Additional Information

A thorough review of BP's medical records revealed that 3 years prior to the current audiological examination, BP presented with a sudden SNHL in his left ear that persisted for 2 months (Fig. 15.2). That audiological examination for the right ear revealed hearing sensitivity to be within normal limits at 250 to 3000 Hz falling to a slight hearing loss at 6000 to 8000 Hz with normal WRS. Results for the left ear, however, revealed normal hearing at 250 to 1000 Hz followed by a moderate to severe SNHL at 1500 to 6000 Hz with recovery to a mild hearing loss at 8000 Hz, and the WRS indicated very poor (8%) recognition for speech. At that time, the patient reported no tinnitus or vertigo and had no history of ototoxic medication. An MRI scan revealed a left cerebellopontine angle (CPA) meningioma (2.4 × 2.3 × 2.4 cm) extending into the left internal auditory canal (IAC) and jugular foramen. The patient underwent Gamma Knife (Elekta, Stockholm, Sweden) stereotactic radiosurgery (GKRS), which reduced the tumor to 1.0 × 1.4 × 1.8 cm. Following GKRS, BP's hearing thresholds and WRS reportedly returned to normal.



AUDIOLOGICAL RECORD

NAME BP

TEST DATE

DOB 3-23-53 SEX Male

AUDIOLOGIST SS

AUDIOMETER

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT				100%	50		
LT				8%	80		

Word Recognition List R ___ L ___ R ___ L ___

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt_____ Lt_____

Peak Pressure (daPa): Rt_____ Lt_____

Type: Rt_____ Lt_____

COMMENTS:

Fig. 15.2 Results from the initial audiological examination.

Two years later, BP perceived slightly decreased hearing. A subsequent audiological examination (Fig. 15.3 indicated hearing to be within normal limits at 250 to 3000 Hz followed by a slight to moderate SNHL at 4000 to 6000 Hz with recovery at 8000 Hz and normal ability to recognize speech (100%). Results for the left ear, on the other hand, revealed normal hearing at 250 Hz followed by a slight to moderate to severe SNHL at 500 to 6000 Hz and recovery to a mild hearing loss at 8000 Hz with normal ability to recognize speech (100%). The SRTs were in agreement with the pure-tone average and indicated a slight loss in the ability to receive speech n the right ear and a mild loss in the left ear. Both ears revealed a "notch" at 6000 Hz that most likely was caused by BP's history of occupational noise exposure. In addition, BP's follow-up MRI scan indicated that BP's tumor was stable and greatly reduced in size compared with the status at earlier audiological examination.

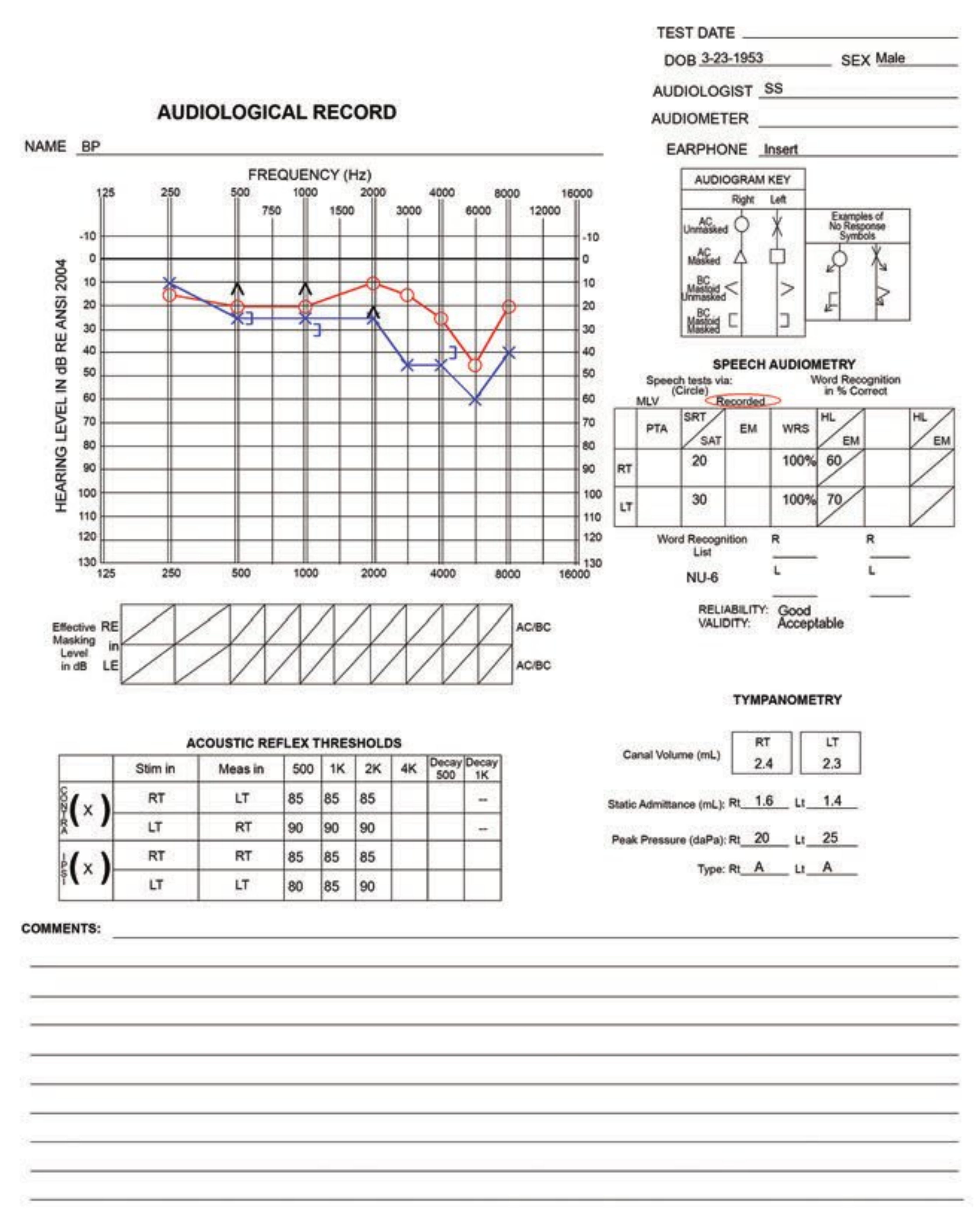
One year later, the patient presented with left upper lung mesothelioma, cisplatin chemotherapy was initiated, and the current audiological examination (Fig. 15.1) was completed.

15.6 Additional Questions to the Reader

1. Given this additional information from the case history, how can the results of the current audiological examination be interpreted?
2. How can the results of the current audiological examination be misinterpreted related to what a clinician might expect from cisplatin therapy?
3. Why would the left ear be more susceptible to chemotherapeutic agents?
4. Was the additional case history information provided by BP necessary to make an accurate diagnosis?

15.7 Discussion of Additional Questions to the Reader

1. Given this additional information from the case history, how can the results of the current audiological examination be interpreted?



NAME BP

TEST DATE

DOB 3-23-1953 SEX Male

AUDIOLOGIST SS

AUDIOMETER

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		20		100%	60		
LT		30		100%	70		

Word Recognition List R L R L

NU-6

RELIABILITY: Good
VALIDITY: Acceptable

Effective Masking Level in dB RE in LE AC/BC AC/BC

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	85	85	85			--
	LT	RT	90	90	90			--
IPSI (X)	RT	RT	85	85	85			
	LT	LT	80	85	90			

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	2.4	2.3

Static Admittance (mL): Rt 1.6 Lt 1.4

Peak Pressure (daPa): Rt 20 Lt 25

Type: Rt A Lt A

COMMENTS:

Fig. 15.3 Results of the audiological examination following Gamma Knife stereotactic radiosurgery.

The components of the current audiological examination that are incongruous with cisplatin ototoxicity are RI and the resulting ART pattern. Both findings are consistent with extra-axial retrocochlear involvement. Review of the audiological examination obtained following GKRS (Fig. 15.3) revealed that WRS and ARTs returned to normal. Thus absence of ARTs with the stimulus presented to the left ear, the significant decrease in WRS inconsistent with the magnitude and configuration of hearing loss, and the presence of RI raise a "red flag" for potential recurrence of BP's CPA tumor. These suspicions were confirmed when an MRI scan following the audiological examination indicated progression of the left meningioma (2.7 × 2.9 × 2.4 cm). Although the tumor had consistently encroached upon the jugular foramen, imaging now revealed compression and deformation of the left brachium pontis (middle cerebellar peduncle) and anterior encroachment of the left IAC.

2. How can the results of the current audiological examination be misinterpreted related to what a clinician might expect from cisplatin therapy?
   BP presented with bilateral hearing loss and tinnitus. Results of audiological examination revealed bilateral asymmetric sensorineural hearing loss with progression primarily in the high frequencies for the right ear (3000–8000 Hz) and 500 to 8000 Hz in the left ear. WRSs decreased in the left ear as might be expected with an asymmetric decrease in hearing thresholds. All these findings are consistent with results commonly observed in cases of cisplatin ototoxicity. Clues to retrocochlear involvement arose from additional test measures that may not be routinely conducted, including abnormal ART pattern and rollover when measuring PB$_{max}$.
3. Why would the left ear be more susceptible to chemotherapeutic agents?
   Prior hearing loss, noise exposure, and radiation are all factors that increase susceptibility to the ototoxic effects of cisplatin. Although both ears demonstrated early signs of noise-induced hearing loss (e.g., 6000 Hz "noise notch"), only the left ear had substantial preexisting hearing loss and exposure to radiation (GKRS).
4. Was the additional case history information provided by BP necessary to make an accurate diagnosis?
   Although having the patient's prior case history would certainly be important for a clinician, the presence of an extra-axial retrocochlear lesion was apparent without knowledge

of the existing diagnosis. Although the patient was referred for possible ototoxicity-induced hearing loss, it was important to identify and react appropriately to incongruous results. The inclusion of ARTs and WRS completed at multiple presentation levels highlighted a change in BP's status beyond expected results that would be exclusively related to ototoxicity.

15.8 Diagnosis and Recommended Treatment

1. Bilateral SNHL secondary to cisplatin ototoxicity and the probable contribution of noise exposure
2. Recurrence of left CPA meningioma with encroachment of left IAC

15.9 Outcome

Results and thorough interpretation were communicated to the referring oncologist treating BP's mesothelioma as well as to the radiation oncologist monitoring BP's meningioma. Cisplatin was replaced with a less ototoxic chemotherapeutic agent due to documentation of hearing loss. Due to the aggressive nature of BP's mesothelioma, treatment for the meningioma was postponed. Unfortunately, BP passed away 7 months later.

15.10 Key Points

1. The clinician must critically analyze all aspects of the audiological assessment from the case history through the audiological examination. Often, outcomes may conflict with anticipated results, and the clinician must react appropriately by modifying the test battery to include appropriate measures and provide a comprehensive detailed interpretation of the results.
2. Results may not be clear and may not be related due to a single pathological contributor. It is important for a clinician not to focus exclusively on the anticipated outcome, but rather to understand that results may be complex and may suggest incorporating additional tests or developing different recommendations.
3. Appropriate interpretation and communication of results to the treatment team is equally if not more important than completing appropriate test measures.

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Part 2

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16 Unilateral Hearing Loss and Disequilibrium

Christopher D. Bauch and Neil T. Shepard

A 20-year-old male was referred for evaluation of unilateral hearing loss (UHL) and disequilibrium following a barotrauma incident and ear surgery.

16.1 Clinical History and Description

TG is a 20-year-old male college student referred for UHL and disequilibrium. He has a complex history of fluctuating right ear mixed hearing loss and vestibular complaints, apparently arising from a poorly described episode 6 years earlier involving barotrauma to the right ear from an air hose in an industrial arts class. TG reported difficulty hearing in the right ear following the trauma.

Two years posttrauma TG experienced otalgia and hearing loss in the right ear. The patient reported that an exploratory surgical procedure was performed in the right ear, perhaps ossiculoplasty, in which the bones were "glued together." Following the procedure, the patient reported a reduction of the otalgia, but no change in hearing.

Three years posttrauma he underwent two audiological examinations 1 month apart at an outside clinic, each indicating normal hearing in the left ear but mixed hearing loss in the right ear ranging from moderate to severe to profound with air–bone gaps between 20 and 55 dB HL. TG also reported sudden onset of persistent vertigo, which was exacerbated by exertion (holding nose and blowing), but resolved within 6 weeks following perilymphatic fistula repair of the right ear.

The following summer (4 years posttrauma) TG sneezed and heard a "pop" in the right ear, resulting in dizziness and unsteadiness worsened by exertion. Outside audiological results again indicated normal hearing in the left ear, but a profound mixed hearing loss in the right ear with 25 to 50 dB HL air–bone gaps. Tympanometry was reported as normal for the left ear with a multiple-peaked tympanogram for the right ear. A second perilymphatic fistula repair was performed, yielding reduced vertigo and gradually resolving unsteadiness.

The next summer (5 years posttrauma) TG experienced three episodes of spontaneous vertigo lasting a few hours to several days, resolving spontaneously and without auditory symptoms. From October to January, TG had persistent motion sensitivity that progressed to mild vertiginous symptoms, causing chronic unsteadiness and the use of a wheelchair. In October, TG also developed symptoms of a headache with associated nausea and reported photophobia with some episodes. TG also reported sudden onset of vertigo lasting 4 or 5 days but gradually improving. Outside audiological testing at that time indicated normal hearing in his left ear, but his right ear revealed a severe to profound (80–100 dB HL) mixed hearing loss with improved bone conduction thresholds resulting in air–bone gaps greater than 60 dB HL, positive Stenger results at multiple frequencies, and a 100% word recognition score (WRS) at 80 dB HL presentation level. Tympanograms were reported as normal bilaterally. Postural control assessment was not evaluated because he was unable to stand unassisted. Rotational chair testing was normal.

At the time of presentation at the current clinic (6 years posttrauma) TG was experiencing general feelings of unsteadiness provoked by walking in grocery store aisles as well as other types of visually provoked motion sensitivity. TG reportedly still had reduced hearing in his right ear. He did obtain a hearing aid for the right ear at an outside clinic but reportedly did not use it.

16.2 Audiological Testing

The patient was evaluated over 3 separate days, with the first clinic visit involving radiology studies with a high-resolution computed tomographic (CT) scan of the temporal bones without contrast. The second visit included conventional audiological and electrophysiological examinations as well as a comprehensive vestibular examination. The third visit included otologic and psychiatric examinations in addition to a physical therapy examination for the Vestibular and Balance Rehabilitation Program.

16.2.1 Radiology Studies

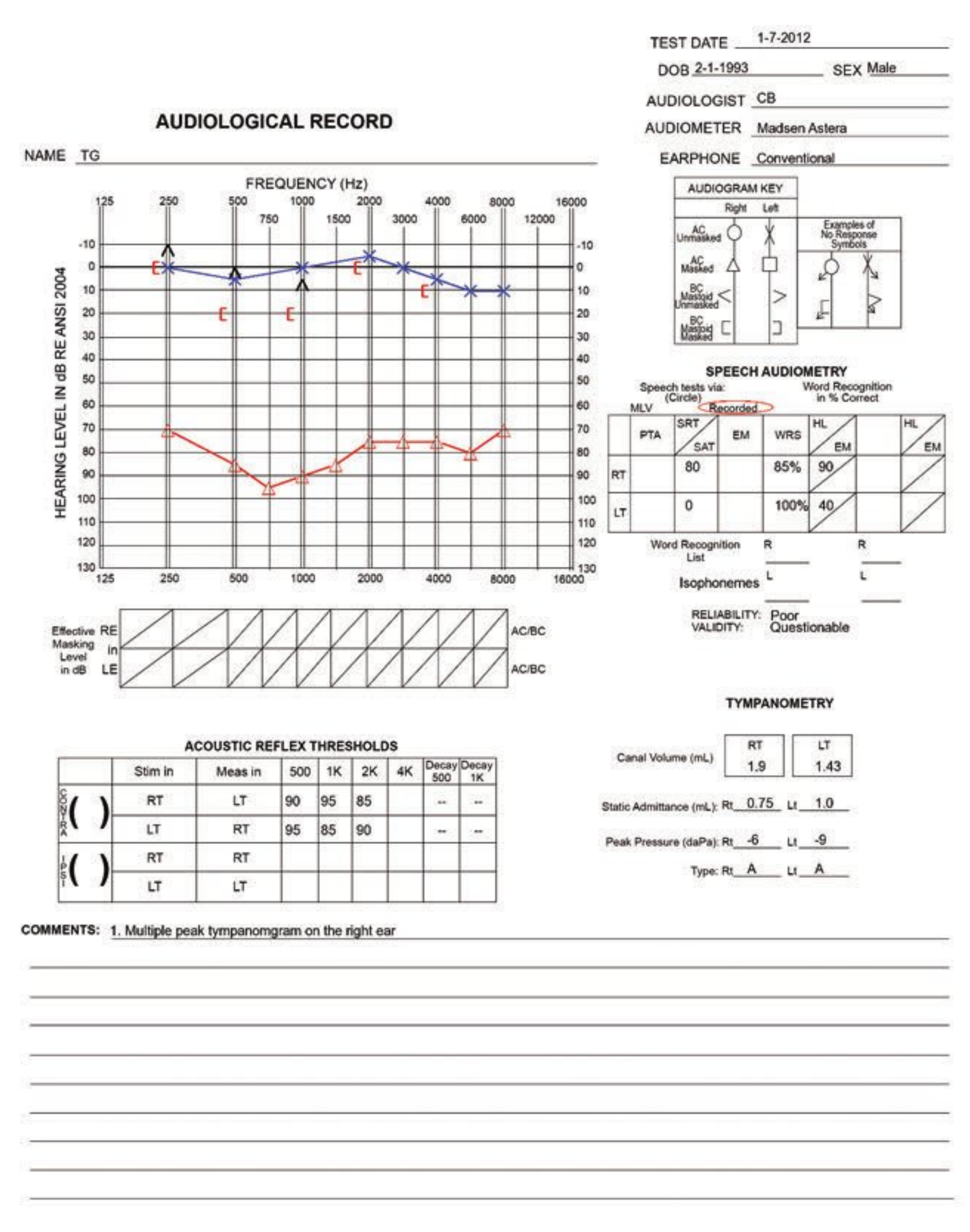
The patient reported he had a normal magnetic resonance imaging (MRI) scan of the brain with and without contrast at an outside clinic. The current CT scan of the posterior fossa/sella/orbit without intravenous (IV) contrast indicated thinning of the roof of the superior canal on the right ear but no definite areas of focal bone dehiscence. Temporal bones were otherwise unremarkable.

16.2.2 Audiological Examination

The results from the initial audiological examination (Fig. 16.1) revealed normal hearing sensitivity for the left ear, but results were inconsistent for the right ear, revealing a unilateral severe conductive hearing loss at all test frequencies with air–bone gaps of 65 to 75 dB HL across frequencies. A masked speech recognition threshold (SRT) was 80 dB HL for the right ear. The masked word recognition score (WRS) in the right ear was 85% when isophonemic 20-word lists were presented at 90 dB HL [10 dB Sensation Level (SL) re: SRT]. Immittance testing indicated normal middle ear pressure and middle ear compliance bilaterally, with a multiple-peaked right ear tympanogram. Contralateral acoustic reflex thresholds (ARTs) were obtained at normal hearing levels bilaterally (85–95 dB HL) despite the apparent severe "conductive" component in the right ear. Pure-tone Stenger testing was positive at 500, 1000, 2000, and 4000 Hz in the right ear.

16.2.3 Otoacoustic Emissions

Distortion-product otoacoustic emissions (DPOAEs) (65/55 dB paradigm) were present for the right ear from 2000 to 4000 Hz, and 2000 to 6000 Hz for the left ear, indicating generally normal outer hair cell function bilaterally.



AUDIOLOGICAL RECORD

NAME TG

TEST DATE 1-7-2012

DOB 2-1-1993 SEX Male

AUDIOLOGIST CB

AUDIOMETER Madsen Astera

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		80		85%	90		
LT		0		100%	40		

Word Recognition List R ___ R ___

Isophonemes L ___ L ___

RELIABILITY: Poor

VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.9	1.43

Static Admittance (mL): Rt 0.75 Lt 1.0

Peak Pressure (daPa): Rt -6 Lt -9

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	90	95	85		--	--
	LT	RT	95	85	90		--	--
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: 1. Multiple peak tympanomgram on the right ear

Fig. 16.1 Initial audiological evaluation indicating normal hearing in the left ear and a severe "conductive" hearing loss in the right ear. Note excellent word recognition scores for the right ear at a reduced sensation level, normal middle ear pressure bilaterally, and normal acoustic reflex thresholds and reflex decay bilaterally. Also noted during examination were positive Stenger results for the right ear at 500 to 4000 Hz.

16.2.4 Auditory Brainstem Response Evaluation

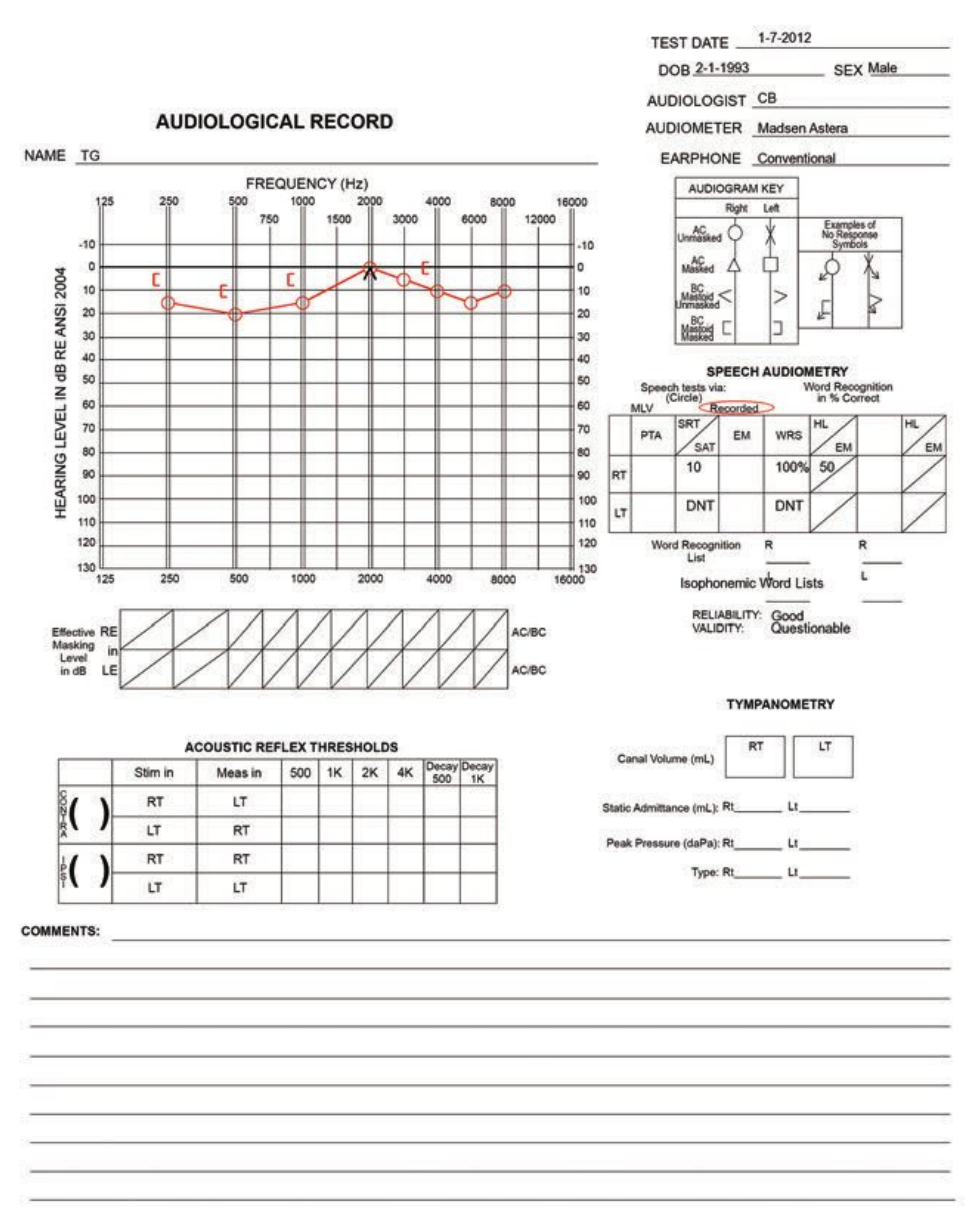
A threshold auditory brainstem response (ABR) evaluation was administered for the right ear only using standard rarefaction click stimuli. Repeatable Wave V responses were easily obtained at appropriate latencies at 20 dB nHL (normalized hearing level), indicating generally normal hearing sensitivity for frequencies between 1000 and 4000 Hz. ABR testing was terminated at this point without further reduction in stimulus intensities.

16.2.5 Clinical Vestibular Evaluation

The clinical vestibular and balance studies were negative for anxiety and depression via the Hospital Anxiety and Depression Scale (HADS). Testing consisted of videonystagmography (VNG) subtests, ocular motor studies, rotary chair, vestibular-evoked myogenic potentials (VEMPs), and dynamic posturography. No abnormal nystagmus was noted with or without visual fixation. Dix-Hallpike and roll tests were negative. Pursuit and saccades were normal. Bithermal caloric irrigation results were normal. Rotational chair results were normal. Ocular and cervical VEMPs were normal on the left and absent on the right, likely secondary to multiple right middle ear surgeries and suspected very mild conductive component on the right. Postural control assessment was normal. Platform pressure tests were all normal. Valsalva procedure against closed glottis and against closed nostrils was negative for symptoms or eye movements. There were no objective indications of either peripheral or central vestibular system involvement noted. He was able to maintain upright stance under a variety of changing sensory input conditions and could coordinate lower limb and upper body reaction to induced forward or backward sway.

16.2.6 Follow-up Audiological Examination

Because of the inconsistencies obtained during the initial hearing evaluation earlier in the day, coupled with the normal findings for ARTs, OAEs, and ABR threshold testing, TG received a repeat right ear audiological examination from a second audiologist (Fig. 16.2). The audiological examination was performed following a frank discussion (with the second author) of how



AUDIOLOGICAL RECORD

NAME TG

TEST DATE 1-7-2012

DOB 2-1-1993 SEX Male

AUDIOLOGIST CB

AUDIOMETER Madsen Astera

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		10		100%	50		
LT		DNT		DNT			

Word Recognition List R ____ R ____

Isophonemic Word Lists L ____ L ____

RELIABILITY: Good
VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt_______ Lt_______

Peak Pressure (daPa): Rt_______ Lt_______

Type: Rt_______ Lt_______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 16.2 Right ear hearing results at the end of the first day indicating normal hearing sensitivity at 250 to 8000 Hz with a slight conductive component at 250 to 1000 and 4000 Hz. Speech recognition threshold and word recognition scores are normal.

the perception of hearing loss could result from a conditioned response over time even though the hearing may have recovered. Given TG's long history of inconsistent right ear results, "pretesting" instructions were provided in a careful and detailed manner. Consequently, reliable and consistent audiological results were obtained for the right ear and indicated normal hearing sensitivity from 250 to 8000 Hz with slight (10 dB HL) air–bone gaps from 250 to 1000 Hz, in addition to normal SRT and WRS.

16.2.7 Otologic Evaluation

Otologic evaluation noted the normal right ear hearing sensitivity following repeat audiological examination, supported by normal ARTs, OAEs, and ABR threshold testing. Otologic examination further indicated some scarring on the right tympanic membrane and tympanosclerosis in the posterior-inferior quadrant of the tympanic membrane. The tympanic membrane was slightly thickened but mobile. The chorda tympani nerve was traversing the posterior-superior quadrant of the tympanic membrane. There was no observation of spontaneous nystagmus. In view of the absence of central or peripheral vestibular abnormalities it was felt chronic subjective dizziness evaluation from the behavioral medicine program for dizziness in psychiatry would be appropriate.

16.2.8 Psychiatric Consultation

Psychiatric consultation included the Mini International Neuropsychiatric Interview (MINI) and found no significant depression or anxiety. Noted was the possibility of vestibular migraine based on the sudden onset of vertiginous symptoms that could persist even when TG was still and the relative absence of aural symptoms accompanied by migrainous features. TG exhibited no evidence of psychoses or suicidality, and no deficits in attention, memory, or concentration. The indications for possible chronic subjective dizziness syndrome were now mostly resolved.

16.2.9 Physical Therapy

The patient reported feeling normal at the time of evaluation with no vestibular/balance symptoms. Vestibulo-ocular reflex (VOR), oculomotor testing, position changes, and gait were all within normal limits. Umbrella optokinetics testing induced a "warm" feeling similar to the grocery store aisle symptoms. Plan of care was to restore his ability to move in grocery store aisles without symptoms using umbrella optokinetics in direct vision via habituation therapy techniques.

16.3 Questions to the Reader

1. Do the presenting symptoms reflect the possibility of a perilymphatic fistula of the round/oval windows, or of the superior canal from a dehiscence?
2. What special audiological testing could be used to help rule out an active fistula?
3. What "nonphysiological" audiological examination results signaled increased suspicion for pseudohypacusis (i.e., functional hearing loss)?
4. Were other "physiological" audiological test measures useful in determining the hearing sensitivity?

16.4 Discussion of Questions to the Reader

1. Do the presenting symptoms reflect the possibility of a perilymphatic fistula of the round/oval windows, or of the superior canal from a dehiscence?
   Yes, the symptoms of mixed fluctuating hearing loss with head movement–provoked dizziness that is intermittent following an antecedent barotraumatic event would fit with a possible recurrent fistula presentation. The segments of TG's presentation that did not fit with fistula (either middle ear or possible canal dehiscence) are the lack of symptoms with exertion and the presentation of spontaneous events of vertigo lasting hours to days that occur without head movements. These latter symptoms would imply another mechanism as the origin for the spontaneous vertigo events beyond a fistula, and given photophobia with the spontaneous events this was likely migraine-related dizziness.
2. What special audiological testing could be used to help rule out an active fistula?
   Of interest was the presenting sign of conductive hearing loss that did not appear of middle ear origin with the normal admittance, which therefore can be suggestive of superior canal dehiscence (SCD). Yet the magnitude of the conductive component was out of character with SCD, and no other objective signs were noted, such as the lack of symptoms with straining and no autophony. The special tests used to rule out an active fistula would be Valsalva against closed glottis and closed nostrils, sealed pneumatic otoscopy, the platform pressure test, and both cervical and ocular vestibular evoked myogenic potentials (cVEMPs and oVEMPs). In the presence of an apparent conductive hearing loss, which may not be completely of middle ear origin if the VEMPs were present, especially with low thresholds and/or elevated amplitudes, then this would be a strong sign for possible SCD. The confounding issue in this case was the past three middle ear surgical procedures; even if the surgical activity did not leave any substantial conductive component it can cause absence of the cVEMP and oVEMP from that side. These tests, in addition to high-resolution CT of the temporal bones, would be needed to determine if an indicated fistula was presently active.
3. What "nonphysiological" test results signaled increased suspicion for pseudohypacusis, (i.e., functional hearing loss)?
   There were a number of "red flags" suggesting unilateral functional hearing loss. Audiological results from the outside clinic and in-house testing indicated inconsistent fluctuating mixed hearing loss in the right ear, ranging from moderate to severe to profound air conduction and shifting bone conduction over time. Inconsistent responses from an otherwise attentive and healthy patient should always alert the tester to the possibility of a functional component. Audiological testing from both clinics also indicated unilateral conductive/mixed hearing loss with air–bone gaps greater than 60 dB HL, which is unacceptably high considering maximal true air–bone gaps should not exceed 60 dB HL. Both clinics reported positive Stenger results for multiple frequencies in the right ear, indicating the patient refused to respond appropriately to audible pure-tone stimuli in the "poorer" ear and providing strong support for functional hearing loss. Further, both clinics reported excellent WRS (85–100%) obtained at a stimulus presentation level equal to, or nearly equal to, the degree of air conduction hearing loss, which is highly unlikely and indicates better hearing sensitivity than obtained.
4. Were other "physiological" audiological test measures useful in determining the hearing sensitivity?
   Yes, there were a number of physiological test measures that helped determine the functional status of the right middle ear, inner ear, and brainstem pathways. Immittance evaluation includes tympanometry and ARTs. Tympanometry assesses middle ear pressure and middle ear compliance, and in this case helped rule out significant middle ear disorder (despite the audiological results indicating significant "mixed" or "conductive" hearing loss). ARTs assess middle ear, cranial nerve VII, and cranial nerve VIII functions and help assess the integrity of the lower brainstem. Importantly, ARTs are often elevated or absent when stimulating an ear with severe or profound hearing loss, or when measuring from an ear with a conductive component. Recall that ARTs were normal bilaterally, indicating bilateral normal middle ear integrity. Otoacoustic emissions (OAEs) require no behavioral responses, provide information on the functional status of the cochlear outer hair cells, and are usually absent with mild or greater sensorineural hearing loss or even slight conductive hearing loss. OAEs are useful in documenting pseudohypacusis. In this case, DPOAE results indicated normal outer hair cell function in the right ear from 2000 to 4000 Hz. Auditory brainstem response (ABR) testing assesses the integrity of CN VIII and ascending brainstem pathways and can provide valuable estimates of hearing levels. In this case, the normal ABR threshold results reflected the best sensitivity in the region between about 1000 and 4000 Hz.

16.5 Diagnosis and Recommended Treatment

Audiological results indicated exaggerated hearing loss in the right ear, which ultimately was determined to be within normal limits with a slight conductive component secondary to previous ossiculoplasty and repairs of perilymphatic fistula. By history, TG had possible perilymphatic fistula, which was inactive at the time of evaluation. TG also had possible migraine-related dizziness. Treatment for the headache component focused on migraine abortive therapies, such as triptans and prochlorperazine. Treatment for the motion-induced dizziness focused on implementing home-based habituation exercises for visual motion sensitivity via umbrella optokinetics in direct vision.

16.6 Outcome

The primary outcome for this patient is to remain in contact with physical therapy should his motion-provoked symptoms become more severe.

16.7 Key Points

1. Audiologists should always be attentive to inconsistent responses during audiological evaluations. Air conduction–bone conduction gaps greater than 60 dB HL and higher than expected WRSs relative to degree of hearing loss are among findings to alert the audiologist to possible functional hearing loss.
2. A number of additional physiological audiological test procedures are useful in determining functional hearing loss, including immittance measures (tympanometry and ARTs), OAEs, and threshold ABR assessment.
3. Vestibular symptoms should be addressed thoroughly. In some cases multiple medical consultations may be needed to provide appropriate diagnosis and treatment.

Suggested Reading

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17 Cochlear Dead Regions (CDR)

Judith Bird and David Baguley

A 74-year-old female attended the hearing clinic on several occasions reporting poor hearing despite consistent use of well-fit digital hearing aids.

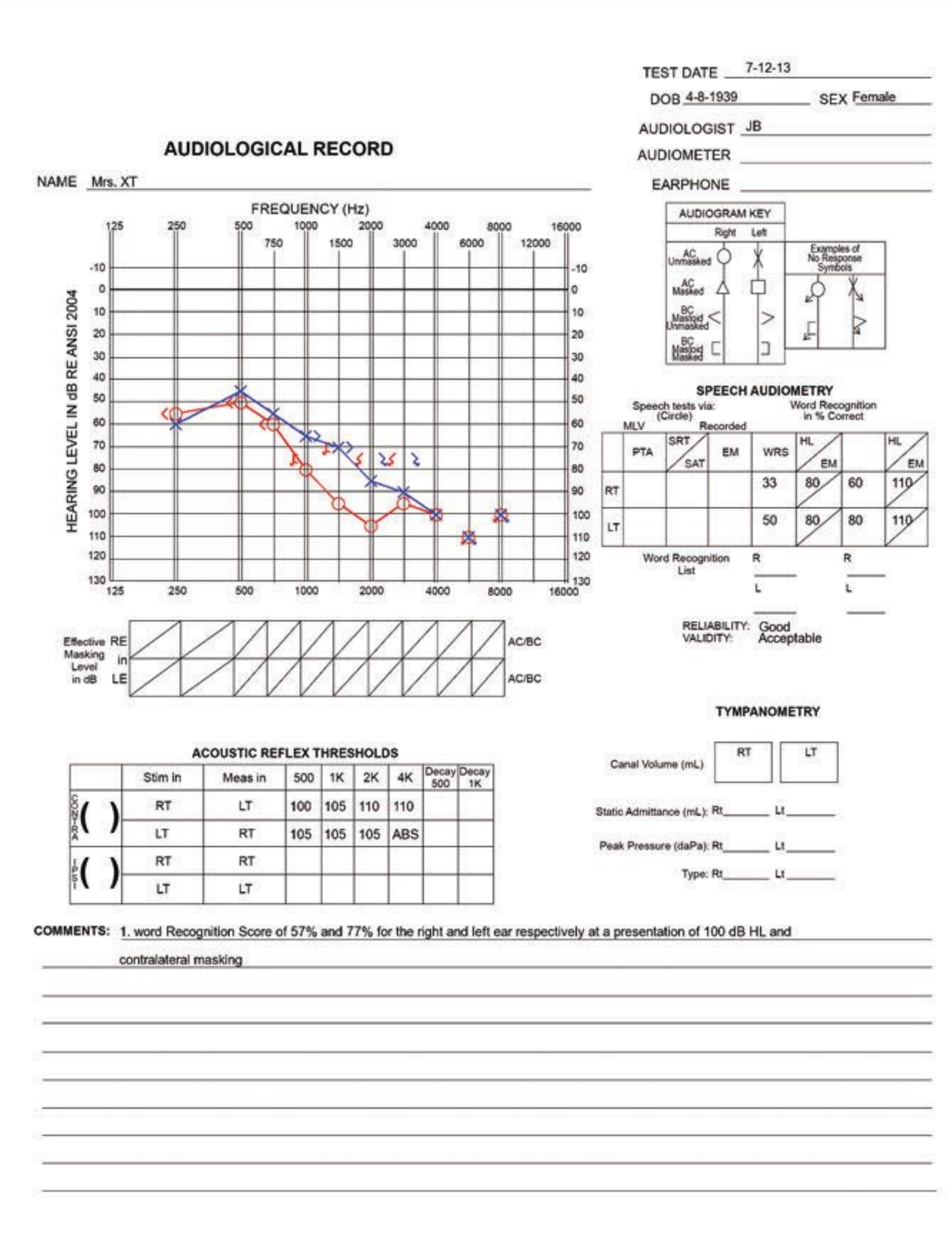
17.1 Clinical History and Description

YT has a long-standing bilateral asymmetrical (right poorer than left at 1000 to 2000 Hz) moderate sensorineural hearing loss sloping to severe/profound sensorineural hearing loss in the high frequencies. She has worn bilateral hearing aids for 5 years but reports that she does not receive the benefit she had hoped for. She reports there is a lack of clarity, particularly when listening in noise. Furthermore, she describes the sound of the aids as tinny and harsh, especially when listening to sounds at high input levels.

YT works as a volunteer for a local charity helping the elderly manage their hearing aids. She has become aware that the benefit she receives from her hearing aids appears to be lower compared to that of the people she encounters. She is concerned that her hearing aids are not programmed correctly. She schedules frequent appointments (nine appointments in the last 2 years) for her hearing aids to be reprogrammed, hoping to achieve better performance.

17.2 Audiological Testing

The results for YT's audiological examination confirmed a bilateral asymmetric sensorineural hearing loss (SNHL) with poorer hearing in the right ear at 1000 to 2000 Hz when compared with the left ear. Results for the right ear revealed a moderate SNHL at 250 to 500 Hz sharply sloping to a profound SNHL at 8000 Hz. Results for the left ear revealed a moderate to severe SNHL at 250 Hz improving to a moderate SNHL at 500 Hz and then sharply sloping to a profound SNHL at 8000 Hz (□ Fig. 17.1). Speech audiometry confirmed very poor ability to


TEST DATE 7-12-13

DOB 4-8-1939 SEX Female

AUDIOLOGIST JB

AUDIOMETER

EARPHONE

AUDIOLOGICAL RECORD

NAME Mrs. XT

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT				33	80	60	110
LT				50	80	80	110

Word Recognition List R ___ L ___ R ___ L ___

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt______ Lt______

Peak Pressure (daPa): Rt______ Lt______

Type: Rt______ Lt______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	100	105	110	110		
	LT	RT	105	105	105	ABS		
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: 1. word Recognition Score of 57% and 77% for the right and left ear respectively at a presentation of 100 dB HL and contralateral masking

Fig. 17.1 Audiogram showing bilateral high-frequency sensorineural hearing loss for YT.

recognize speech bilaterally with results for the right ear poorer than the left ear. YT was tested for cochlear dead regions (CDRs) using the threshold equalizing noise (TEN) test. During this test, the listener has to respond to tones with and without the presence of the threshold equalizing noise (TEN) in the same ear. A positive result for the presence of a CDR is indicated when thresholds in the presence of the TEN are at least 10 dB greater than the same threshold measured without noise and the level of the noise. It is assumed that when the intensity of the tone is increased, the excitation spreads along the basilar membrane, and the stimulus is detected at a location with functioning inner hair cells (IHCs). For YT, such “off-frequency listening” was found to be present when presenting a 1000 and 3000 Hz tone to the right ear and at 3000 Hz tone to the left ear. This finding suggests the presence of CDRs in those frequency regions for the right and left ears. The TEN test could not be completed at all frequencies due to the magnitude of the hearing loss.

17.3 Questions to the Reader

1. What are CDRs and how prevalent are they?
2. What is the value of testing for CDRs, and should all candidates for hearing aids be tested using the TEN test?
3. What is the evidence to suggest that the hearing aid gain-frequency response should be altered to reflect the presence of CDRs?
4. How can the information about CDRs be used for optimizing patient management?

17.4 Discussion of Questions to the Reader

1. What are CDRs and how prevalent are they?
   CDR refers to areas of the basilar membrane where there is a complete loss of IHC function. Prevalence estimates vary depending on the population sample studied. Using the conventional TEN test criteria (thresholds in the presence of the TEN are are at least 10 dB greater than threshold measured without the presence of noise and the level of the noise), estimates range from 33%for patients with moderate to severe hearing loss to 57%for patients with mild to severe hearing loss having CDRs for at least one test frequency.
2. What is the value of testing for CDRs and should all candidates for hearing aids be tested using the TEN test?
   CDRs are rare for hearing thresholds 60 dB HL. Therefore, unless there is a clear indication of poor word recognition or a patient reports speech and/or sound to be distorted, performing the TEN test in patients with hearing thresholds better than 60 dB HL is unlikely to be of value. In those patients with severe hearing loss, administering the TEN test can be difficult at higher levels due to loudness discomfort.

   The value of testing for CDRs has been debated. Most current prescription formulae, such as National Acoustic Laboratories’ nonlinear fitting formula version 1 (NAL-NL1), assume reduced cochlear function at poorer hearing thresholds,. This has been labeled “hearing loss desensitization.” Hence there is less prescribed gain when hearing thresholds exceed 70 to 80 dB HL. Routine testing using the TEN test for all patients is therefore probably not indicated. Although testing for CDRs can be done quickly in the clinic, its value is greatest for patients with severe hearing loss (e.g., 70 dB HL or greater), patients reporting distortion, or patients with particular difficulty in word recognition.
3. What is the evidence to suggest that the hearing aid gain-frequency response should be altered to reflect the presence of CDRs?
   Studies have shown that patients with CDRs in the high frequencies obtain less benefit from high-frequency amplification than those who do not have CDRs. This seems to be particularly true where there are CDRs at more than one frequency. There is some evidence to suggest that providing amplification where CDRs exist can actually reduce user performance. Where high-frequency CDRs have been identified, Baer et al suggested that sound is amplified up to 1.7 times the edge frequency (the lowest frequency where the CDR is measured). For low-frequency CDRs, 0.57 times the edge frequency (in this case, the highest frequency where a CDR was found to be present) has been recommended.

   Conversely, it has been suggested that identification of one or two high-frequency dead regions does not require gain modification because some studies have not reported a detrimental effect.
4. How can the information about CDRs be used for optimizing patient management?
   If the addition of amplification in the presence of a CDR reduces speech recognition then reducing the gain is clearly indicated. For patients where increasing audibility by maximizing high-frequency gain is not providing benefit, there may be some secondary benefits of reducing unnecessary gain. For example, this may provide reduced feedback or improve listening comfort. Furthermore, having objective correlates of described difficulties can aid counseling by increasing the understanding of the limitations of amplification. This can help redirect therapy toward hearing strategies such as optimal positioning and reducing background noise, speechreading, and other assistive listening devices (e.g. remote wireless microphones that stream to the hearing aids) that may improve the signal-to-noise ratio (SNR).

   TEN test results can also be used as a basis for referral for cochlear implantation (CI). There have been examples in the literature where patients with apparently usable hearing at moderate threshold levels have been shown to have significant CDRs and ultimately proceeded to CI. Results from the TEN test can provide additional information to supplement and reinforce the results from conventional speech audiometry and help confirm insufficient benefit from hearing aids.

17.5 Diagnosis and Recommended Treatment

Testing for CDRs in YT was indicated by the documented very poor speech recognition scores. Given the presence of significant CDRs in the right ear, the amplification for the right hearing aid was reduced above 1700 Hz. This eased YT’s report of loudness discomfort as well as the presence of feedback. Counseling was provided about other communication strategies such as optimal positioning to reduce the impact of background noise as well as. YT's attending speechreading classes.

Testing was completed to assess word recognition ability, and aided speech tests indicated performance of 73%(sentences presented at 65 dBA using monitored live voice without visual cues). This score is better than the current UK criteria for CI of 50% word recognition in quiet without the use of visual cues. YT's hearing will continue to be monitored at least every 3 years, and a decision to refer for CI will be reviewed.

17.6 Outcome

The test results gave the audiologist greater confidence in reducing high-frequency amplification where otherwise there might be concern of reduced audibility. This reduction in amplification above 1700 Hz in the right ear resulted in greater listening comfort as YT had found the high-frequency sound tinny and unpleasant. In this case, however, the primary value of having the additional information about CDR provided by the results from the TEN test was in that it provided better counseling about YT's realistic expectations from amplification. Given the results of the TEN test, YT came to understand that there were underlying reasons to explain the additional difficulty she was experiencing compared with her peers. Once having accepted this, YT was more receptive to counseling and training about communication tactics, assistive devices, and speechreading. She also explored using a remote wireless microphone to improve SNR for use in meetings. Her satisfaction with her hearing aids improved, and further appointments were not requested over the subsequent 2 years.

17.7 Key Points

1. Unusually poor speech recognition abilities warrant further investigation.
2. The TEN test is a useful tool to further investigate decreases in speech recognition abilities and can indicate the presence of CDRs. The TEN test is easy to administer and interpret.
3. A positive TEN test warrants careful review of hearing aid settings. Decreasing amplification that is not improving speech recognition may yield indirect benefits in listening comfort or reduction of feedback. In rare cases, speech recognition scores have been shown to improve by reducing hearing aid gain at frequencies within the measured CDR.
4. Information about CDRs improves counseling by helping to create realistic expectations from hearing aid performance and highlights the need to explore other strategies to improve communication, such as reducing background noise, optimal positioning, and speechreading.
5. Time invested in fully understanding the hearing needs of a patient will increase efficient use of resources by reducing repeated visits to the clinic.

Suggested Reading

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18 ABR versus MRI in the Diagnosis of Acoustic Tumors

Alison M. Brockmeyer-Lauer

The two patients discussed in this case report, patient A and patient B, presented with asymmetric hearing loss, and auditory brainstem response (ABR) results suggested the presence of auditory nerve or auditory brainstem dysfunction. Magnetic resonance imaging (MRI) confirmed the presence of an acoustic tumor, specifically a vestibular schwannoma, in patient A, but patient B's MRI scan was normal.

18.1 Clinical History and Description

18.1.1 Patient A

Patient A is a 58-year-old female who first scheduled an appointment with an otologist for medical evaluation of asymmetric hearing loss and imbalance. She reported slow, progressive bilateral hearing loss over the past 10 to 15 years. Approximately 2 years ago, however, she experienced a sudden onset of vertigo, lasting several days. Following the vertigo episode, the hearing in the right ear decreased rapidly, and she reported the hearing in the right ear was significantly poorer than in the left ear. She also stated she can no longer recognize speech in the right ear. She has not had any additional episodes of vertigo, but reports significant difficulty with balance. The hearing in her left ear has not significantly decreased over the last 2 years, but she has noticed fluctuations in the hearing in her left ear associated with intermittent aural fullness. She reports intermittent tinnitus in the right ear, which occurs frequently throughout the day. She denies otalgia, and excessive noise exposure, as well as aural fullness in the right ear. She reports a familial history of age-related and noise-induced hearing loss in her father. She currently uses left bilateral contralateral routing of signal (BICROS) amplification, which she finds very helpful. Finally, magnetic resonance imaging (MRI) was ordered by the original referring physician. When the films were reviewed by the consulting otologist, he was concerned that there was a small lesion in the right internal auditory canal (IAC). He felt, however, that the films were less than optimal.

18.1.2 Patient B

Patient B is a 76-year-old male who scheduled an appointment for an audiological examination due to decreased hearing. He reported difficulty hearing his wife and other women's voices. He also noted a change in the quality of music, specifically of treble sounds. He stated he often attends the theater and has difficulty in certain listening environments with poor acoustics or poor pronunciation. He feels his hearing is symmetric and states that he uses the telephone on the left ear due to habit. He reports long-standing bilateral tinnitus, but denies dizziness, otalgia, and aural fullness. He also denies excessive noise exposure but reports a familial history of hearing loss in his maternal grandfather.

18.2 Audiological Testing

18.2.1 Patient A

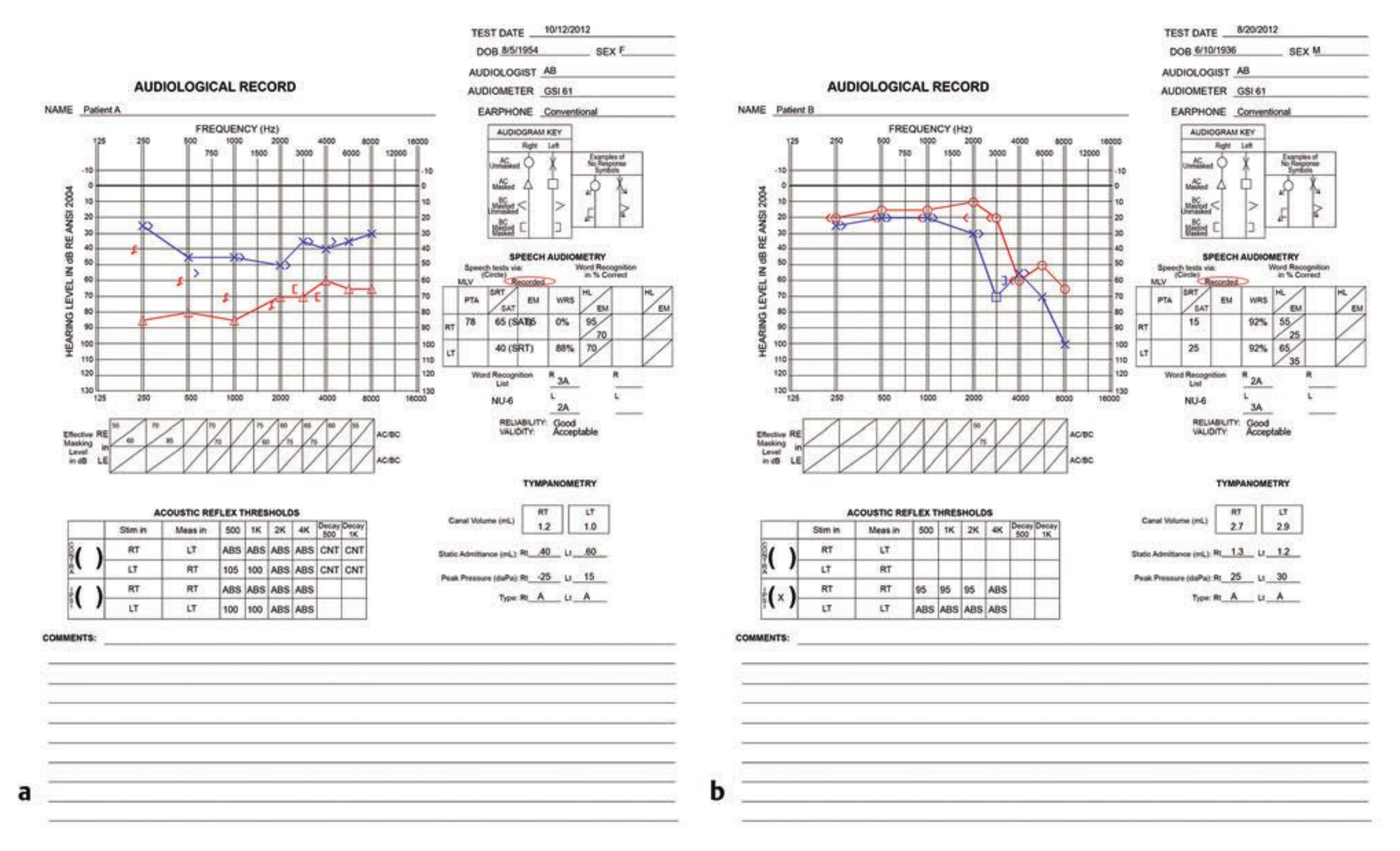
Audiological examination for the left ear revealed a slight sensorineural hearing loss (SNHL) at 250 Hz, sloping to a moderate SNHL from 500 to 2000 Hz, rising to a mild SNHL from 3000 to 8000 Hz. The right ear presented with severe SNHL from 250 to 1000 Hz, rising to moderately severe SNHL from 2000 to 8000 Hz (□ Fig. 18.1a). Speech recognition threshold (SRT) for the left ear revealed a mild loss in the ability to recognize speech. A speech awareness threshold (SAT) was obtained in the right ear, which revealed a moderately severe loss in the ability to be aware of the presence of speech. Word recognition scores (WRSs) were obtained using a recorded version of Northwestern University Auditory Test No. 6 (NU-6) monosyllabic word lists with a female speaker. Words were presented at most intelligible level (MIL), corresponding to a 30 dB sensation level (dB SL) relative to the SRT and SAT. A score of 88% was obtained in the left ear, demonstrating slight difficulty in word recognition ability. A score of 0% was obtained in the right ear, demonstrating very poor word recognition ability. Immittance audiometry was performed, and the tympanograms were within normal limits bilaterally. With stimulation to the left ear, ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present but elevated at 500 and 1000 Hz and absent at 2000 and 4000 Hz. Ipsilateral and contralateral ARTs with stimulation in the right ear were absent 500 to 4000 Hz. Due to the asymmetric hearing loss, it was recommended that Patient A follow up with the otologist and retrocochlear pathology be ruled out. The otologist then ordered an ABR test. A second MRI scan was also ordered because the otologist felt that the original MRI films were less than optimal.

Patient A returned for ABR testing. ABR testing was completed using rarefaction and alternating polarity click stimuli presented at 11.1 and 67.1 clicks per second (□ Fig. 18.2a). Stimulus presentation levels of 75 and 94 dBnHL were used in the left and right ears, respectively. Technical recording quality was good. ABR results revealed normal absolute latencies for Waves I, III, and V on the left side and normal interpeak latencies for Waves I to III, III to V, and I to V on the left side. Also, a normal shift in Wave V latency was observed with increased stimulus repetition rate on the left side. All waveforms on the right side were absent, suggesting auditory nerve or auditory brainstem dysfunction on the right side.

A second MRI scan of the brain and brainstem was performed (□ Fig. 18.3a), and the otologist noted a lesion in the right IAC. It was determined that the right IAC was filled with an enhancing mass in the superior and inferior compartments, but the mass did not extend into the cerebellopontine angle.

18.2.2 Patient B

Audiological examination for the right ear revealed a slight SNHL at 250 Hz, rising to normal hearing sensitivity from 500



a

AUDIOLOGICAL RECORD

NAME Patient A

TEST DATE 10/12/2012

DOB 8/5/1954 SEX F

AUDIOLOGIST AB

AUDIOMETER GSI 61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	HL / EM
RT	78	65 (SAT)	65	0%	95 / 70	
LT		40 (SRT)		88%	70	

Word Recognition List NU-6: R 3A; L 2A

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	105	100	ABS	ABS	CNT	CNT
IPSI ( )	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	100	100	ABS	ABS		

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.2	1.0

Static Admittance (mL): Rt .40 Lt .60

Peak Pressure (daPa): Rt -25 Lt 15

Type: Rt A Lt A

COMMENTS:

b

AUDIOLOGICAL RECORD

NAME Patient B

TEST DATE 8/20/2012

DOB 6/10/1936 SEX M

AUDIOLOGIST AB

AUDIOMETER GSI 61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	HL / EM
RT		15		92%	55 / 25	
LT		25		92%	65 / 35	

Word Recognition List NU-6: R 2A; L 3A

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI (x)	RT	RT	95	95	95	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	2.7	2.9

Static Admittance (mL): Rt 1.3 Lt 1.2

Peak Pressure (daPa): Rt 25 Lt 30

Type: Rt A Lt A

COMMENTS:

Fig. 18.1 Recent audiological examinations for patient A (a) and patient B (b).

to 2000 Hz and then sloping to a moderately severe SNHL at 4000 and 8000 Hz. The left ear presented with a slight SNHL from 250 to 1000 Hz, sloping from mild SNHL at 2000 Hz to a profound SNHL at 8000 Hz. A significant asymmetry was present at 2000, 3000, 6000, and 8000 Hz (Fig. 18.1b). The SRT for the right ear was normal, and the SRT for the left ear revealed a slight loss in the ability to recognize speech. WRSs were obtained using a recorded version of NU-6 monosyllabic word lists with a female speaker. Words were presented at MIL, corresponding to a 40 dB SL relative to the SRT. A score of 92% was obtained bilaterally, demonstrating normal ability to recognize speech. Immittance audiometry revealed tympanograms that were within normal limits bilaterally. Ipsilateral ARTs in the right ear were present within normal limits 500 to 2000 Hz, and absent at 4000 Hz. Ipsilateral ARTs in the left ear were absent 500 to 4000 Hz. Due to the asymmetric hearing loss, it was recommended that Patient B follow up with an otologist and retrocochlear pathology be ruled out. The otologist then ordered an ABR test.

Patient B returned for ABR testing, which was completed using rarefaction and alternating polarity click stimuli presented at 11.1 and 67.1 clicks per second (Fig. 18.2b). Stimulus presentation level of 90 dBnHL was used in the right ear, and stimulus presentation levels of 90 and 94 dBnHL were used in the left ear. Technical recording quality was fair. ABR test results showed normal absolute latencies for Waves I, III, and V on the right side and normal interpeak latencies for Waves I to III, III to V, and I to V on the right side. A slightly prolonged shift in Wave V latency was observed with increased stimulus repetition rate on the right side. ABR test results on the left side showed normal absolute latency for Wave I and delayed absolute latency for Wave V. Wave III was absent on the left side. A prolonged I to V interpeak latency was observed on the left side and increased stimulus repetition rate was not performed on the left side. ABR test results suggested auditory nerve or auditory brainstem dysfunction on the left side.

MRI of the brain and brainstem was ordered by his otologist following the ABR to rule out a tumor on the auditory nerve. The MRI scan was normal (Fig. 18.3b).

18.3 Questions to the Reader

1. Both patients had abnormal ABR test results, but only patient A had an acoustic tumor based on the results of the MRI scan. How do ABR and MRI differ in the diagnosis of acoustic tumors?
2. If MRI is the gold standard in the diagnosis of acoustic tumors, what role does otoneurological ABR testing play in diagnostic audiology?
3. Why is accurate diagnosis of hearing loss site of lesion important?

18.4 Discussion of Questions to the Reader

1. Both patients had abnormal ABR test results, but only patient A had an acoustic tumor based on the results of the MRI scan. How do ABR and MRI differ in the diagnosis of acoustic tumors?

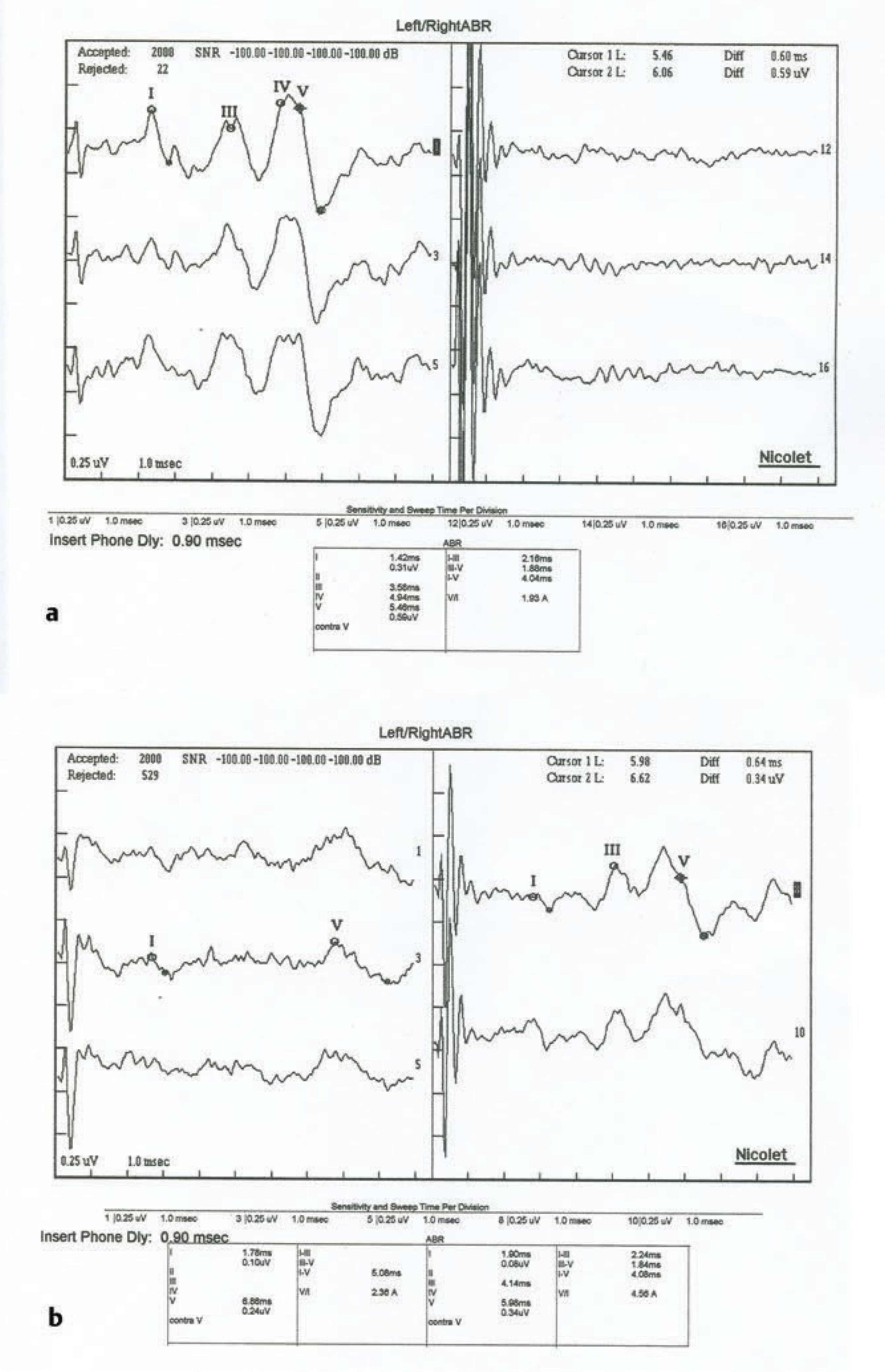


Fig. 18.2 Auditory brainstem response waveforms for patient A (a) and patient B (b).

The ABR, when first introduced to audiology, became very popular because it was a sensitive test for the identification of acoustic tumors. Advances in MRI techniques, however, led to even greater sensitivity in the identification of acoustic tumors. MRI allows for the identification of small tissue masses, and it quickly became the gold standard in identifying acoustic tumors. The ABR, however, continues to suggest auditory nerve or auditory brainstem dysfunction in approximately 90% of the cases with acoustic tumors that are greater than 1.5 cm, whereas the ABR only reports abnormalities approximately 70% of the time with tumors smaller than 1.5 cm.

It is important, however, to remember that the ABR and MRI are different tests. The ABR is a test that evaluates auditory function, whereas the MRI scan assesses anatomical structures. Auditory dysfunction is not always related to structural changes as seen with MRI. The ABR can detect auditory dysfunction caused by infection, metabolic disease, or degenerative disease, which are not detected by MRI. As a result, it is possible to observe abnormal ABR test results with a normal MRI scan, as was seen with patient B in this case report.

2. If MRI is the gold standard in the diagnosis of acoustic tumors, what role does otoneurological ABR testing play in diagnostic audiology?
   As discussed in Question 1, abnormal ABR test results indicate auditory nerve or auditory brainstem dysfunction and can assist in the diagnosis of the hearing loss site of lesion. In

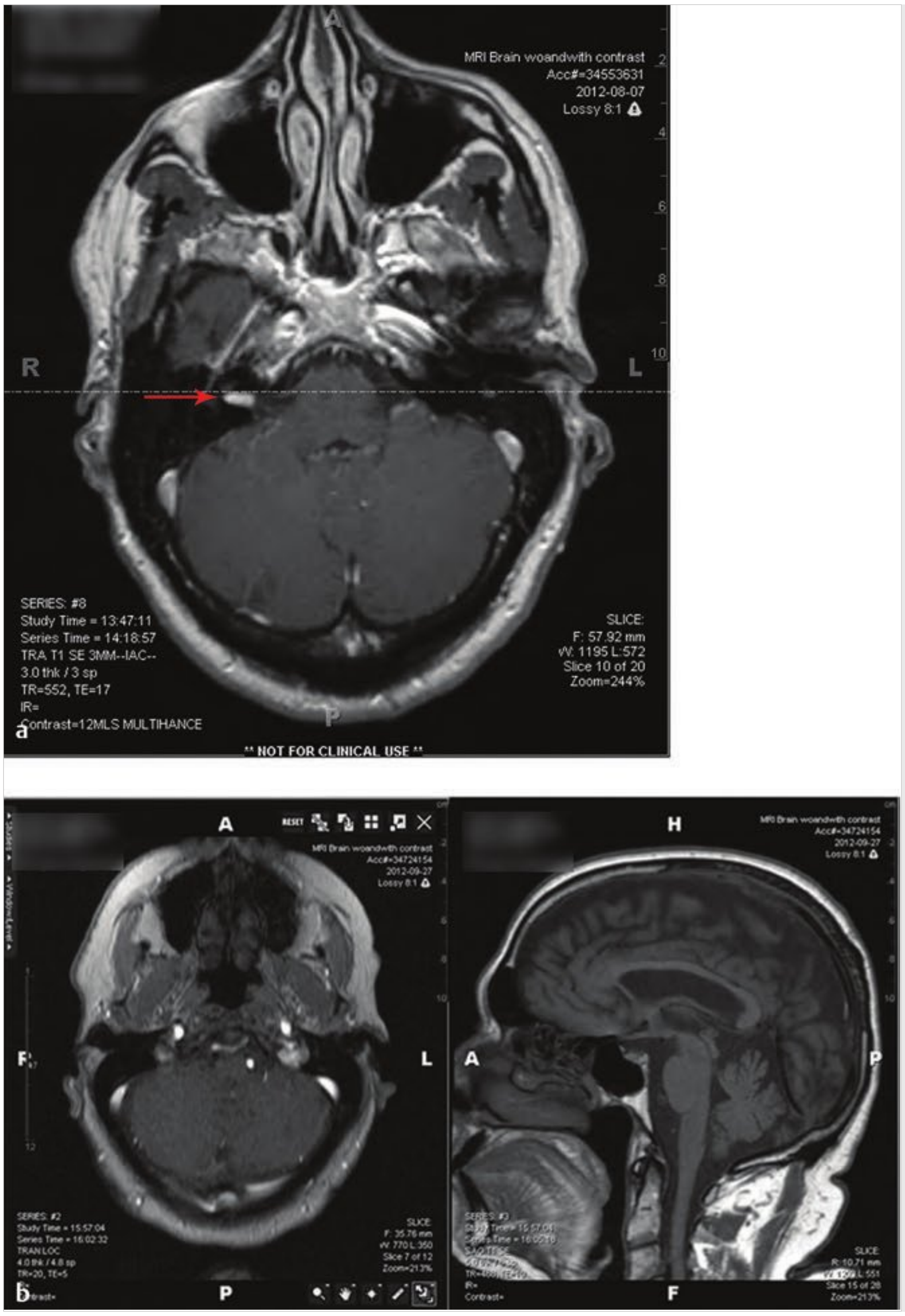


Fig. 18.3 Magnetic resonance imaging scans for patient A (a) and patient B (b). The lesion on the right side in Patient A's scan is highlighted with a red arrow.

the case of Patient B, it could easily be assumed that the hearing loss in the left ear is cochlear due to the normal MRI scan. The absence, however, of all left ARTs is not consistent with cochlear loss. The abnormal ABR on the left side confirms a neural component to the hearing loss. If only the MRI scan had been performed, then the hearing loss in the left ear would likely have been assumed to be cochlear. However, Patient B's hearing loss in the left ear likely has a neural component not caused by structural changes seen with MRI. Proper diagnosis of site of lesion requires a battery of tests, and diagnostic evaluation should continue even if an acoustic tumor is ruled out by MRI. MRI is the gold standard in the diagnosis of acoustic tumors, but ABR continues to be the preferred test in the evaluation of neural function.

3. Why is accurate diagnosis of hearing loss site of lesion important?

   Accurate diagnosis of hearing loss site of lesion can be very important for appropriate patient counseling and treatment. Neural hearing loss often leads to poorer word recognition ability than would be expected based on pure-tone thresholds. When a neural site of lesion is identified, the patient can be counseled on realistic expectations from amplification, and it can be explained that amplification cannot restore sound clarity. In the case of patient B, the word recognition ability in the left ear is within normal limits. Because ABR results revealed auditory nerve or auditory brainstem dysfunction on the left side, however, Patient B can be counseled that word recognition in the left ear may be greatly affected if the hearing loss progresses.

   Also, treatment of neural hearing loss may include the recommendation not only of amplification but also of hearing assistive technologies (HATs), such as personal FM systems. HATs can improve the patient's ability to receive and recognize speech and other auditory signals compared to performance with amplification alone. Finally, if the patient has poorer speech recognition in one ear due to neural hearing loss, then special devices, such as BICROS amplification, may be recommended. Patient A uses a left wireless BICROS device. With the BICROS device, the sound is routed from a microphone transmitter on her poorer right ear to a hearing aid receiver on her better left ear. Her poorer right ear cannot use a traditional hearing aid due to the very poor word recognition ability.

18.5 Diagnosis and Recommended Treatment

Patient A was diagnosed with an acoustic tumor, specifically a right solitary vestibular schwannoma. A solitary vestibular schwannoma is a single tumor that originates in the vestibular portion of the eighth cranial nerve, just within the internal auditory canal. Patient A already uses a left BICROS hearing aid to improve her communication, and it was recommended that she continue its use. She has significant hearing loss in her better-hearing left ear and is unable to wear a conventional hearing aid in her poorer right ear due to her very poor word recognition ability. The left BICROS hearing aid provides the needed amplification in the left ear, while also routing sound from her poorer right ear to the better left ear. HATs, including a personally worn FM system and an amplified phone, were also recommended for patient A. She was encouraged to return for annual audiological examinations and to use hearing protection in noise to protect the usable hearing in her better left ear. Finally, patient A followed-up with her otologist regarding treatment and monitoring of the vestibular schwannoma as well as for management of her imbalance.

Patient B was diagnosed with asymmetric SNHL and was medically cleared for amplification if desired. It was also recommended that he return for a repeat audiological examination in 1 year and that he use hearing protection in noisy environments.

18.6 Outcome

Patient A successfully uses a left BICROS hearing aid and follows up with her audiologist for maintenance and programming. The vestibular schwannoma is being monitored by her otologist with annual MRI scans. She is, however, experiencing more balance difficulty than expected for the small size of the tumor. Vestibular examination revealed hypofunction of the right vestibular system. She has received two intratympanic gentamicin injections in an attempt to stabilize the right hypofunction to allow for central compensation. If significant imbalance continues, the otologist is recommending complete surgical resection of the schwannoma.

Patient B has decided not to pursue amplification at this time. Although he noticed difficulty hearing in certain listening situations, he does not find the difficulties to be significant enough to pursue amplification. He is scheduled for an annual audiological examination to monitor his hearing and the asymmetry.

18.7 Key Points

1. ABR and MRI are different diagnostic tools. The ABR is a test that measures auditory function, whereas MRI evaluates anatomical structure.
2. The ABR can reveal auditory dysfunction secondary to infection, degenerative disease, and metabolic disorders, which cannot be diagnosed through MRI.
3. The ABR is an important tool in the diagnostic test battery and when used in conjunction with other diagnostic tools, ABR can facilitate accurate diagnosis of hearing loss site of lesion.
4. Proper diagnosis of hearing loss site of lesion can be very helpful in counseling the patient on realistic expectations and can guide the recommended treatment.

Suggested Reading

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19 My Hearing Aids Do Not Help Me at All

Lauren Harvey and Wayne J. Wilson

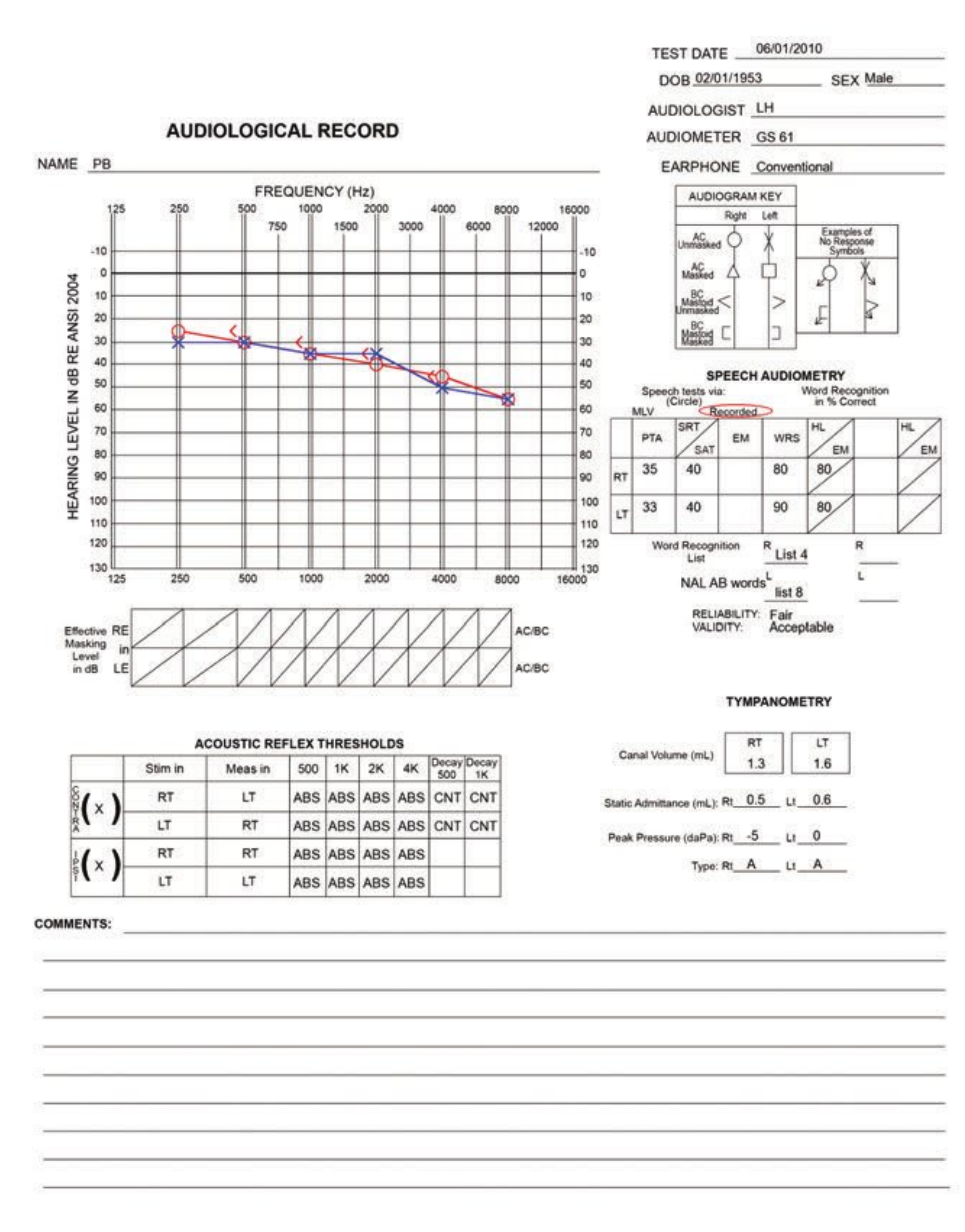
A 57-year-old male was referred by his audiologist for further diagnostic assessment as the patient neared the end of a surprisingly unsuccessful trial of hearing aids.

19.1 Clinical History and Description

PB is a 57-year-old male who has been on a 4-week trial of bilateral behind-the-ear (BTE) hearing aids after self-referring to his audiologist. PB originally presented to his audiologist seeking an audiological examination and a hearing aid trial. PB stated that his hearing had been slowly deteriorating over the past 5 years to the point where he was having difficulty hearing in most listening environments and particularly in noisy listening environments. This was starting to negatively affect PB's work as an accountant where he regularly attended meetings with groups of clients and talked to clients over the phone. It was also starting to affect PB's home life where he often had to look after his "quickly speaking" grandchildren, and his need to increase the volume on the television was becoming a source of agitation between him and his wife. PB also reported infrequently hearing a "buzzing noise" in his head; no history of vertigo, noise exposure, or hearing loss in the family; and no significant medical events other than taking over-the-counter medication for hay fever.

19.2 Audiological Testing

Fig. 19.1 reports the results of PB's initial audiological examination performed by his audiologist. Pure-tone testing revealed bilateral symmetrical mild-to-moderate sensorineural hearing loss (SNHL) that is gradually falling in configuration. Because this testing was completed in Australia, PB's word recognition scores (WRSs) were obtained using the National Acoustics



TEST DATE 06/01/2010

DOB 02/01/1953 SEX Male

AUDIOLOGIST LH

AUDIOMETER GS 61

EARPHONE Conventional

NAME PB

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	35	40		80	80		
LT	33	40		90	80		

Word Recognition List R List 4 R

NAL AB words L list 8 L

RELIABILITY: Fair

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.3	1.6

Static Admittance (mL): Rt 0.5 Lt 0.6

Peak Pressure (daPa): Rt -5 Lt 0

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (X)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS:

Fig. 19.1 PB's audiological examination.

Laboratories' Arthur Boothroyd (NAL-AB) word lists and their recommended protocols. The NAL-AB word lists contain 15 lists of 10 monosyllabic words. The patient's response to each word is scored phonemically (30 scoreable phonemes per list) such that correctly repeating three, two, one, or none of the phonemes in each word elicits a score of 10%, 7%, 3% or 0% for that word, respectively. A performance-intensity (PI) function is obtained for each ear by presenting the first word list at the expected half-maximum level (HML: the level at which the patient is predicted to score 50%) and subsequent lists at levels 15 dB higher than the previous list until a maximum score is reached. If not already found for the first word list, a score is also obtained at a signal level below HML. Speech noise was used for masking and was applied to the nontest ear according to the following formula: masking level in nontest ear = presentation level in test ear - 40 dB + air-bone gap in nontest ear + audiometer conversion factor. Once the PI function is plotted, the SRT is estimated by extrapolating the presentation level required to reach the 50% score on the PI function. PB's speech recognition thresholds (SRTs) revealed a mild loss in the ability to receive speech bilaterally. PB's WRSs revealed a slight loss in the ability to recognize speech in the right ear and normal ability to recognize speech in the left. PB's WRSs improved with increased presentation levels to a maximum of 80% in the right ear and 90% in the left ear at the 80 dB HL presentation level with no significant rollover at a 90 dB HL presentation level.

Tympanometry revealed middle ear pressure (daPa), static compliance (mL), and ear canal volume (mL) to be within the normal range; however, PB's acoustic reflex thresholds (ARTs) were absent to ipsilateral and contralateral stimuli at 500 to 4000 Hz.

19.3 Diagnosis and Recommended Treatment

In light of PB's case history and audiological results, PB's audiologist recommended that PB consider a 4-week trial of hearing aids. The audiologist also recommended PB make an appointment to see his family physician to seek medical clearance to trial hearing aids and a medical opinion on his absent ARTs. PB agreed to both recommendations. PB secured medical clearance from his family physician who also requested the audiologist to monitor PB's hearing (particularly his ARTs) on a 6-monthly basis. PB then returned to the audiologist for a hearing aid fitting. The audiologist fit PB with bilateral BTE hearing aids to National Acoustics Laboratories prescriptive procedure for fitting non-linear hearing aids, version 2 (NAL-NL2) real ear insertion gain (REIG) targets, counseled PB on how to use his new hearing aids, and started PB on his 4-week trial.

Three weeks into his 4-week hearing aid trial, PB returned to his audiologist complaining that, although his hearing aids were of some benefit in quiet listening environments, his hearing aids were of no benefit and often made things worse in noisy listening environments. When asked to elaborate, PB replied that it was as if his hearing aids mix noise with the voices PB is trying to hear. When this happens, PB reported having to remove his hearing aids to hear what is being said. The audiologist reassessed PB's hearing and revealed no change from PB's previous assessment. The audiologist also assessed PB's hearing aids in a hearing aid analyzer and revealed PB's hearing aids were performing within manufacturer specifications. Finally, PB's audiologist also reassessed PB's REIG results and revealed these results still met NAL-NL2 REIG targets.

To further investigate PB's reports, the audiologist used the speech perception in noise (SPIN) test to assess PB's unaided and aided performance when listening to sentences in quiet and noise. The SPIN test assesses final word recognition in sentences with controlled word predictability. In this case, the audiologist presented the SPIN using live-voice at a conversational level to PB as he listened binaurally. Each SPIN list contains 50 sentences of five to eight words in length with each sentence ending in a target word that is predictable in 50% of the sentences and unpredictable in the remaining 50% of the sentences. PB was asked to repeat the final word in each sentence to measure his high predictability (HP) score (the number of final words correctly repeated in sentences where the final word was predictable from the remainder of the sentence) and his low predictability (LP) score (the number of final words correctly repeated in sentences where the final word was not predictable from the remainder of the sentence). In quiet, PB's HP score was 80% and his LP score was 60% when unaided. His HP score was 96% and his LP score was 80% when aided. In the presence of competing 12-talker babble presented from a loudspeaker at 65 dB SPL, PB's HP score was 54% and his LP score was 40% when unaided. His HP score was 18% and his LP score was 6% when aided.

19.4 Question to the Reader

1. Considering PB's complaints regarding the performance of his hearing aids, and the resulting assessments conducted by PB's audiologist to investigate these complaints, what should PB's audiologist do next?

19.5 Discussion of the Question to the Reader

1. Considering PB's complaints regarding the performance of his hearing aids, and the resulting assessments conducted by PB's audiologist in response to these complaints, what should PB's audiologist do next?
   PB's poor results in the first 3 weeks of his hearing aid trial were surprising considering that his initial audiological results suggested he was a good candidate for a hearing aid trial because he had self-reported hearing difficulties and was motivated to try hearing aids. Also, PB's audiological results revealed a bilateral symmetrical, gradually sloping, high-frequency SNHL that was within a range that could be aided, and his WRSs were shown to improve with increased presentation levels bilaterally without any sign of rollover. Also, PB's audiologist was able to program PB's hearing aids to meet NAL-NL2 REIG targets. Of concern, however, was PB's absent ARTs and his hearing aids providing little benefit and often making listening worse in noisy listening environments. This latter concern was supported by PB revealing his poorest scores on the SPIN test in the aided/noise condition. Although PB's audiologist made several programming changes to PB's hearing aids to improve performance in

noise, his audiologist also referred PB to the audiology clinic at a nearby university. This referral requested a comprehensive audiological site of lesion assessment to investigate PB's absent ARTs and poor aided performance when listening in noise compared with his aided performance in quiet.

19.6 Additional Testing

On arriving at the audiology clinic at a nearby university, PB's initial audiological examination revealed no significant change in hearing status. As part of the new examination, PB's transiently evoked otoacoustic emissions (TEOAEs) and distortion-product otoacoustic emissions (DPOAEs) revealed abnormally large amplitudes bilaterally. This suggested a disinhibition (a reduced inhibition) of outer hair cell activity, possibly because of reduced activity in the neural loop that includes afferent cochlear nerve fibers connecting inner hair cells to the brainstem, and efferent cochlear nerve fibers (the olivocochlear bundles) connecting the brainstem to the outer hair cells.

To further investigate PB's reports of performing more poorly in noisy listening environments, PB was assessed using a behavioral central auditory processing disorder (CAPD) test battery based on the recommendations of the American Speech-Language-Hearing Association (2005), and screened using a test of short-term and working memory. This assessment was not conducted to confirm or deny the presence of a CAPD (as any such decision would be confounded by PB's SNHL). Instead, it was conducted to quantify PB's performance in poor listening environments. The CAPD test battery included a low-pass filtered speech (LPFS) test, a competing sentences (CS) test, a two-pair dichotic digits (TPDD) test, a frequency pattern (FP) test, a 500 Hz masking level differences (MLD) test, and the random Gap Detection Test (RGDT), several of which were applied in a nonstandard format:

a) Low-pass filtered speech (LPFS): For each ear separately, PB was asked to repeat 50 monosyllabic words from equivalent word lists presented at 70 dB HL to the test ear. Half the words were filtered (standard format) and half the words were not filtered (nonstandard format). The words were from the Auditec of St. Louis recordings of the Northwestern University Auditory Test No. 6 (NU-6) word lists 1C and 2C and had been recorded using a male talker. The filtered words had been low-pass filtered using a 1000 Hz cutoff frequency.

b) Competing sentences (CS): For each ear separately, PB was asked to repeat 20 sentences presented at 70 dB HL to the test ear. Half the sentences were presented while equivalent competing sentences were simultaneously presented at 70 dB HL to the nontest ear (standard format—dichotic), and half the sentences were presented without these competing sentences (nonstandard format—monotic). The sentences were from the Auditec of St. Louis recordings that were six to seven words in length and had been recorded from a male talker.

c) Two-pair dichotic digits (TPDD): For both ears simultaneously, PB was asked to repeat 20 sets of four digits. Each set of four digits was presented sequentially as two digit pairs with one digit in each pair presented at 70 dB HL (dial setting) to the right ear while the other digit in that pair was simultaneously presented at 70 dB HL (dial setting) to the left ear (standard format—dichotic). For each ear separately, PB was then asked to repeat another 20 sets of four digits where each set of four digits was also presented sequentially as two digit pairs, but each digit in each pair was presented at 70 dB HL to the right ear only (10 sets of four digits) or to the left ear only (10 sets of four digits) (nonstandard format—monotic). The digits were from the U.S. Department of Veterans Affairs recordings that included the numbers 1 through 10 (except 7) and had been recorded using a male talker.

d) Frequency pattern (FP): For each ear separately, PB was asked to describe the frequency patterns contained in 25 sets of three tones presented monotically at 70 dB HL. The frequency patterns were from the U.S. Department of Veterans Affairs recording with each tone in each set of three tones being either 880 Hz or 1,122 Hz.

e) 500 Hz masking level difference (MLD): For both ears simultaneously, PB was asked to indicate if he heard a 500 Hz tone presented in the presence of a 500 Hz narrow-band noise (NBN). This tone-in-noise condition was presented under two test conditions. In the first condition, the tone and noise were in phase (homophasic) with the noise presented at 70 dB HL and the tone reduced in 2 dB steps from a starting signal to noise (SNR) of 1 dB until PB could no longer hear the tone. In the second condition, the tone and noise were 180° out of phase (antiphasic) with the noise presented at 70 dB HL and the tone reduced in 2 dB steps from starting SNR of − 7 dB until PB could no longer hear the tone. The difference between the tonal detection threshold obtained in the homophasic and antiphasic conditions was defined as the MLD.

f) The Random Gap Detection Test (RGDT): For both ears simultaneously, PB was asked to indicate if he had heard one or two tones where the tones were separated by silent gaps of 0, 2, 5, 10, 15, 20, 25, 30, or 40 msec Four series of tone pairs were presented with all tones in each series being 500, 1000, 2000, or 4000 Hz and being presented at 70 dB HL.

The tests of short-term and working memory were four subtests from the Test of Auditory Perceptual Skills—Revised. The first subtest was Auditory Number Memory Forward (ANMF), which tested PB's ability to repeat increasing series of numbers in the order the numbers were spoken. The second subtest was Auditory Number Memory Backward (ANMB), which tested PB's ability to repeat increasing series of numbers in the reverse order the numbers were spoken. The third subtest was Auditory Word Memory (AWM), which tested PB's ability to repeat increasing series of unrelated words not necessarily in the order they were spoken. The final subtest was Auditory Sentence Memory (ASM), which tested PB's ability to repeat increasing-length sentences exactly as they were spoken. The audiologist used monitored live-voice to present these stimuli at a comfortable listening level.

Fig. 19.2 reports PB's results following the LPFS, CS, TPDD, and FP tests from the behavioral (C)APD assessment battery described above. PB also performed poorly on the remaining tests in the CAPD assessment battery, the RGDT and the MLD test. On the RGDT, PB was unable to detect the presence of two tones in any of this test's stimuli. On the 500 Hz MLD test, PB obtained no release from masking on presentation of the 500 Hz tone and 500 Hz NBN in the antiphasic condition. That

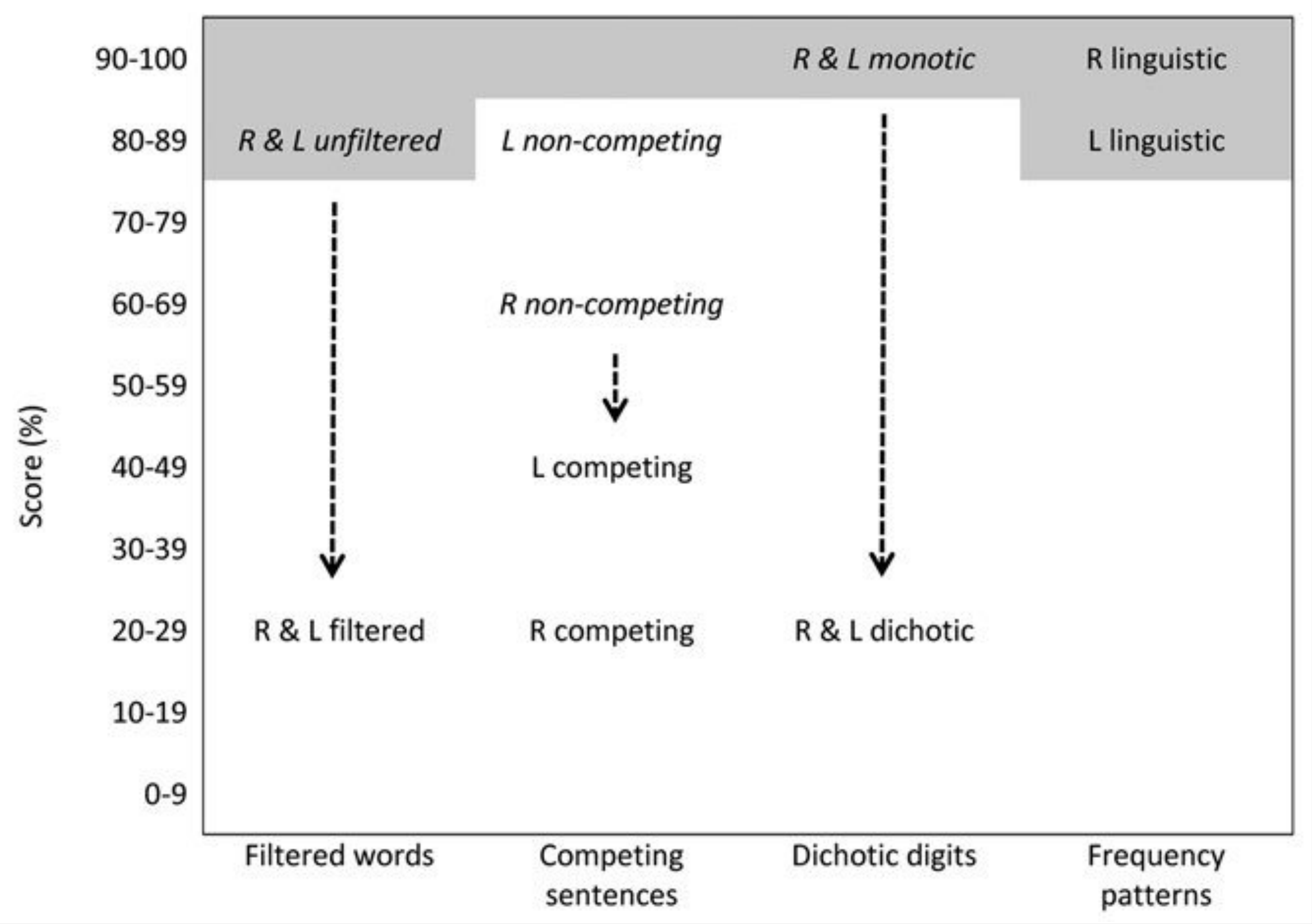


Fig. 19.2 PB s right (R) and left (L) ear scores for four tests from the behavioural central auditory processing disorder (C)APD test battery. The regular text represents the results obtained using the standard test protocols (with the shading representing the respective normal ranges [mean ± 2SD]). The italicized text represents the results obtained using the nonstandard test protocols.

is, PB's performance did not improve from the homophasic to the antiphasic condition. Finally, PB performed well on all the screening tests of tests of short-term and working memory.

In light of PB's continued absence of ARTs, abnormally large amplitude TEOAEs and DPOAEs, and his poor performance on five out of six behavioural CAPD tests, PB was evaluated using the auditory brainstem response (ABR) and the auditory late latency response (ALLR). For the ABR, click stimuli were presented to PB's right and left ears at 90 decibels above normal adult hearing level (dB nHL) at 7.1 clicks per second (cps). For the ALLR, 1000 Hz tone-burst stimuli were presented to PB's right and left ears at 90 dB nHL at 1.1 tone-bursts per second. For the ABR and the ALLR, PB's responses were recorded using an ipsilateral electrode array where the noninverting electrode was placed on vertex, the inverting electrode was placed on the mastoid of the test ear, and the ground electrode was placed on the mastoid of the nontest ear.

Fig. 19.3 reports PB's ABR and ALLR test results following the assessment previously described. PB's ABR results for both ears to alternating click stimuli revealed significantly degraded waveform morphology with no repeatable Wave I and questionable Waves III and V (particularly for the left ear) with estimated absolute and interwave latencies within normal limits. PB's ABR results to the separated rarefaction and condensating click stimuli showed similarly degraded waveforms. This was particularly so for the right ear where no repeatable waves were identified. This result cast further doubt on the reliability of the previous ABR waveforms obtained to alternating click stimuli in the right ear. No evidence of especially prominent cochlear microphonic (CM) activity was noted in any of PB's ABR waveforms to the rarefacting or condensating click stimuli. PB's ALLR results bilaterally revealed degraded waveform morphology and poor repeatability, but waves N1 and P1 were identified at latencies and amplitudes within normal limits.

19.7 Additional Question to the Reader

1. What do the additional audiological tests tell the audiologist about PB's hearing loss?

19.8 Discussion of the Additional Question to the Reader

1. What do the additional audiological tests tell the audiologist about PB's hearing loss?
   The results of the additional audiological tests for PB were considered in two parts: behavioral and objective. Although PB's behavioral CAPD test results were interpreted cautiously because of his SNHL, these results revealed seven primary findings.
   1. PB's ability to repeat words decreased significantly when the words were degraded (LPFS).
   2. PB's ability to repeat sentences decreased significantly when a competing sentence was presented to the other ear (CS).
   3. PB's ability to repeat digits decreased significantly if he had to repeat different digits presented to each ear simultaneously (TPDD).
   4. PB was unable to detect silent gaps of as much as 40 msec between tones (RGDT).
   5. PB was unable to use phase cues to better hear a 500 Hz tone in NBN (MLD).
   6. PB was able to describe in words tonal patterns consisting of three tones (FP).
   7. PB passed a screening assessment of his short-term and working memory.

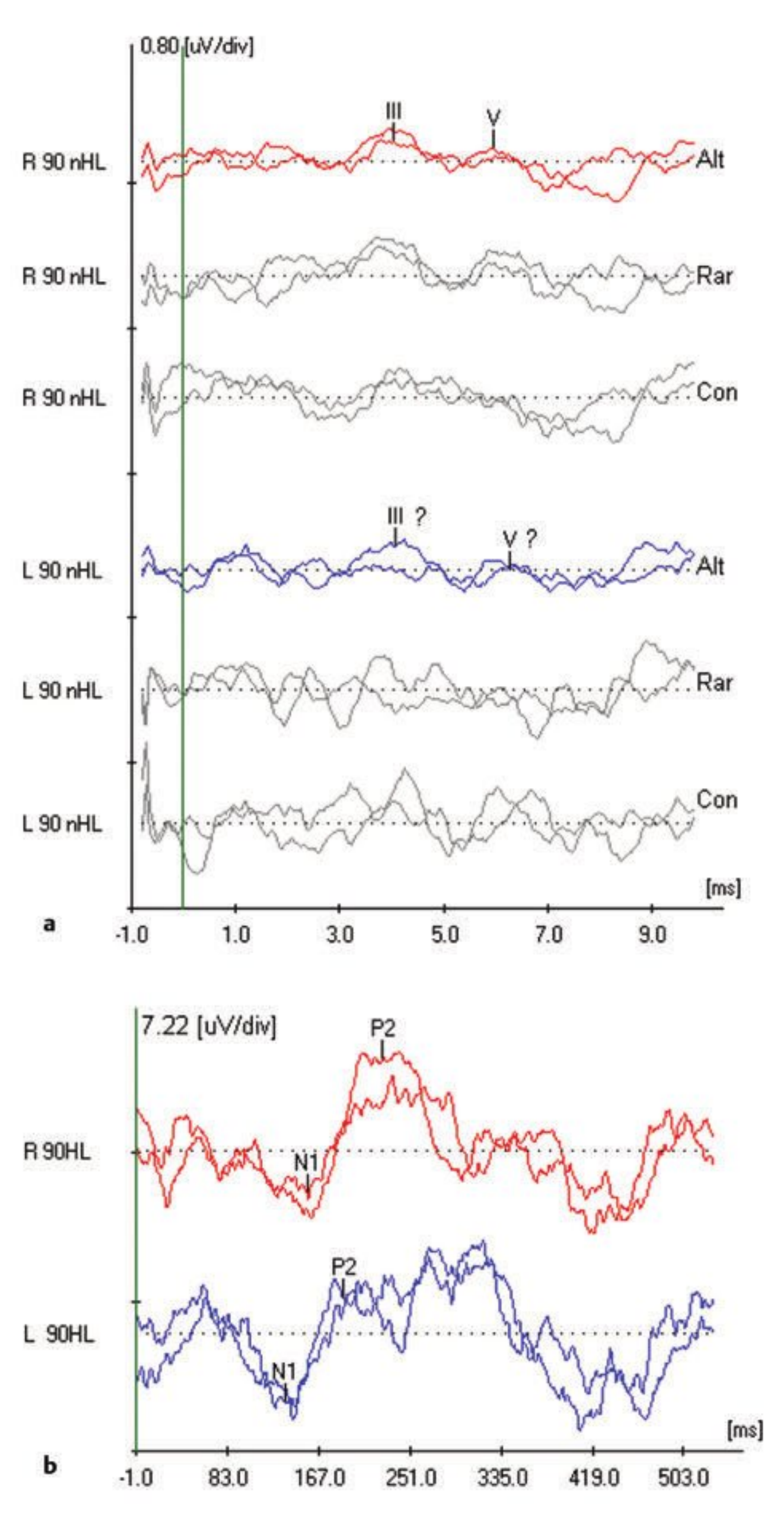


Fig. 19.3 PB's auditory brainstem response and auditory late latency response waveforms. Alt. alternating stimulus; Rar, rarefacting stimulus; Con, condensating stimulus.

From a functional perspective, these results support PB's reports of increased difficulty listening in noisy listening environments. From a site of lesion perspective, and despite the confounding effect of PBs SNHL, the poor performance on five of the seven behavioral tests warranted further audiological investigation of PB's auditory nervous system.

PB's objective audiological test results proved to be of greater diagnostic value because these results were considered to be consistent with auditory neuropathy spectrum disorder (ANSD). This conclusion was drawn from PB's ARTs being absent, his TEOAEs and DPOAEs being present and abnormally large in amplitude, his ABR waveforms being significantly degraded or absent (although these ABR waveforms did not contain especially prominent CM activity), and his ALLR waveforms being degraded.

Berlin et al describe ANSD as a condition in which a person's OAEs and/or cochlear microphonics are, or were at one time. present, and ABRs are abnormal or absent. Berlin et al note that persons with ANSD can have hearing thresholds ranging from within normal limits to a profound hearing loss that can fluctuate. These patients typically have poor speech recognition, particularly when listening in noisy environments. Finally, these patients almost always have absent ARTs.

While still debated, ANSD is thought to be part of a continuum involving varying degrees of dysfunction of the cochlear inner hair cells, the inner hair cell–cochlear nerve fiber junctions, and/or the cochlear nerve fibers. This dysfunction is thought to be related to dissynchronous cochlear nerve fiber activity that distorts the sound information carried on those fibers, especially under conditions where synchronous neural activity is particularly important such as listening in noisy environments. The specific cause(s) of ANSD remain unknown and are the topic of much research with possibilities including neonatal anoxia, hyperbilirubinemia, mutations in otoferlin or pejvakin genes, Charcot-Marie-Tooth Disease, Mohr-Tranebjaerg syndrome, and mitochondrial diseases. It was not known if PB had any of these disorders.

ANSD remains a difficult disorder to treat. Of the available audiological treatment options, hearing aids have generally been unsuccessful. Although a positive relationship has been reported between ALLR waveforms and hearing aid outcomes in children with ANSD, such an outcome was not observed for PB. Cochlear implantation has proven to be a more successful, although still not highly successful, treatment option. Factors such as cost and predicting the likelihood of success for this option remain a challenge. In light of these challenges, nonauditory communication strategies (e.g., visual language approaches such as sign language or cued speech) remain as a treatment alternative.

19.9 Diagnosis and Recommended Treatment

PB's additional audiological testing revealed results consistent with bilateral ANSD. When receiving this information, PB's audiologist referred PB to an otologist and to a neurologist for a comprehensive otoneurological assessment. The audiologist also counseled PB on the high probability that he would continue to receive limited benefit from his hearing aids, especially in noisy environments. Also, PB was counseled that he should consider returning his hearing aids for credit and undergo communication training to learn nonauditory strategies for hearing better in noisy listening environments. Finally, PB was counseled on cochlear implants as a possible treatment option for ANSD. After considering his options, PB agreed to complete the otoneurological assessment and to return to his audiologist for communication training. Surprisingly, PB also elected to continue using his hearing aids as he reported gaining sufficient benefit from his hearing aids in quiet listening environments.

He now removes his hearing aids in noisy listening environments.

19.10 Key Points

1. Although PB's initial audiological assessment suggested he was a good candidate to trial hearing aids, his poor outcomes 3 weeks into his trial contributed to a decision to conduct further audiological assessments. These assessments led to a diagnosis of ANSD that may have otherwise gone undiagnosed.
2. By completing both behavioral and objective assessments, the audiologist was able to better manage PB by offering a more complete audiological diagnosis and a fuller description of the environments in which PB could expect to benefit or not benefit from his hearing aids.

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20 In Utero and Early Childhood Drug Exposure: An Auditory, Speech, and Language Outcome

Robert W. Keith

Intrauterine and early drug exposure may have serious lifelong consequences, including multiple comorbid problems related to memory, attention, reading, speed of processing, language, and audition. The results of an auditory processing disorder (APD) test battery confirmed some of the auditory complaints described in the intake case history and provided clues to a generalized neurological pathway disorder that was inefficient for listening and language function. In spite of these problems, this patient is a highly educated and successful professional. Audiologists should be judicious when counseling patients about possible future achievements in life in the face of significant sensory and cognitive factors.

20.1 Clinical History and Description

NM is a 25-year-old female with difficulty understanding in noise and other degraded speech conditions. She reports a delay in auditory processing that may require “a minute” to process what is being said; in addition, she reports having poor short-term memory. She states she functions well because she has adapted communication strategies to overcome her deficits.

NM’s mother was a drug user for the entire pregnancy, which included opiates, acid, marijuana, and cocaine. She was born via cesarean section due to her mother’s genital herpes. Her APGAR scores were low, and she was placed in the neonatal intensive care unit (NICU) for several months following her birth. As a child, her parents had her use the drugs they were using. NM lived with her birth parents until she was adopted by her babysitter’s family at 8 years of age. Her otologic medical history includes multiple ear infections as a child.

NM attended public schools through fifth grade, when she began to fail. Her mother thought she was “fooling around during school and not paying attention.” NM reports she struggled to understand what was occurring in the classroom. While math was the “easier” subject for NM, “English was a disaster.” She struggled with social studies, science, and all subjects requiring a “heavy” language load. Her phonological skills were poor, and she was a rote sight reader.

In fifth grade, NM was moved to a private school that used an Accelerated Christian Education (ACE) curriculum, which emphasized individualized and nongraded studies allowing students to work at their own level of achievement. The learning environment is extremely quiet; talking is not allowed without permission except for asking a question of the teacher. NM thrived under the ACE program, achieving A’s. In the sixth grade, she attended a public school once again, where she began to fail, and in seventh grade she returned to the ACE program where her grades returned to A’s. NM remained in that program until graduation. In college, NM sat in the front row and avoided large lecture halls, where she always performed poorly.

NM received no formal intervention or remediation at any time while in school. According to NM, the factors that helped were “the quiet environment, the forced reading comprehension, and the learning on my own pace and each section building on a basic concept from the previous day.” Currently, NM continues to have many of the reading, language, and listening problems as described earlier. When NM is extremely tired she becomes more frustrated and does not cope as well, especially when she knows she is struggling. In spite of these factors, NM has earned a doctoral degree and works with children having disabilities.

20.2 Audiological Testing

The APD test battery included pure-tone and speech audiometry, the SCAN-3A Tests for Auditory Processing Disorders (APDs) in Adolescents and Adults (Table 20.1), Staggered Spondee Word Test (SSW), Dichotic Digits Test (DDT) presenting double digits using free recall response instructions, and the BKB SIN Speech-in-Noise Test (Etymotic Research, Elk Grove Village, IL, 2002). Otoacoustic emissions (OAEs), acoustic reflex thresholds (ARTs), reflex decay, and auditory brainstem responses (ABRs) were obtained at another center, and the results were forwarded for review.

Results from pure-tone and speech audiometry revealed hearing to be within normal limits bilaterally and NM’s word recognition scores (WRSs) were 100% OAEs were present, ARTs were elevated, and reflex decay was present bilaterally. The ABR was reported to be abnormal, requiring a 90 dB nHL signal to obtain a repeatable response. The neurotologist who reviewed the findings diagnosed NM as having a “rare form of auditory neuropathy.”

Results of the APD test battery are summarized in Table 20.2, Table 20.3, Table 20.4, Table 20.5, Table 20.6, and a description of the subtests of the SCAN-3A test battery is provided following here.

In addition to the results reported in these tables, the results from the BKB-SIN test revealed a signal to noise ratio (SNR) loss of 8.5 dB, indicating “moderate” difficulty understanding sentences in the presence of background noise. The core tests of the SCAN-3A test battery reported in Table 20.2 indicate performance ranging from 1 to 25%, and NM’s total composite score was in the 2nd percentile for persons in NM’s age group. Results of the supplementary tests of the SCAN-3A are reported in Table 20.3, and NM’s performance ranged from 1st to the 53rd percentile with the Time Compressed Sentences (TCS) test performance substantially above all other test results. Comparison of the Competing Words–Directed Ear (CW-DE) (25th percentile) to the Competing Words–Free Recall (CW-FR) (37th percentile) finds the standard score and percentile to be essentially equal. This comparison indicates that there is no cognitive or attention factor confounding the findings, and the results are indicative of abnormal auditory processing abilities.

Inspection of ear advantage was calculated by subtracting the left ear (LE) from the right ear (RE) as reported in Table 20.4.

Table 20.1 SCAN-3:A Tests for Auditory Processing Disorders for Adolescents and Adults administered to the subject of this case report

Gap Detection	This screening test measures a patient's ability to detect brief silent gaps of variable durations (measured in milliseconds) between tone pairs.
Auditory Figure-Ground (AFG) + 8 dB	This test is used to assess a patient's ability to understand speech in the presence of background noise at a + 8 dB signal to noise ratio (i.e., the stimulus words are 8 dB of greater intensity than the background multitalker speech). This test is used as a screening test and/or a diagnostic test.
Competing Words–Free Recall (CW-FR)	This test is used to assess the patient's ability to understand competing speech signals by presenting a monosyllabic word pair simultaneously to each ear. The patient is instructed to repeat both words in any order. This test is used as a screening test and/or a supplementary test
Filtered Words (FW)	This diagnostic test is used to assess the patient's ability to understand distorted speech by presenting monosyllabic words low-pass filtered at 750 Hz.
Competing Words–Directed Ear (CW-DE-RE; CW-DE-LE)	This diagnostic test is used to assess the patient's ability to understand competing speech signals by presenting a monosyllabic word pair simultaneously to the left and right ears. The patient is instructed to repeat both words in a prescribed order.
Competing Sentences (CS)	This diagnostic test is used to assess the patient's ability to understand competing speech signals by presenting pairs of unrelated sentences to the right and left ears. The patient is instructed to repeat the sentence heard in one ear and ignore the sentence heard in the other ear.
Auditory Figure-Ground 0 dB, + 12 dB (AFG 0; AFG 12)	These tests are used to assess the patient's ability to understand speech in the presence of background noise, at the same intensity as the background noise (0 dB) and at a + 12 dB signal to noise ratio (i.e., the stimulus words are of 12 dB greater intensity than the background multitalker speech).
Time Compressed Sentences (TCS)	This supplementary test is used to assess the patient's ability to process degraded speech by presenting sentences that have been time compressed at 60%

Table 20.2 SCAN:3A Tests for Auditory Processing Disorders test and composite scores

Age	Raw score	Standard score	Confidence interval	Percentile rank
Filtered Word	22	6	4–8	9%
Auditory Figure-Ground (AFG)	21	5	3–8	5%
Competing Words (CW)	43	8	6–10	25%
Competing Sentences (CS)	53	3	2–4	1%
Composite score		69	64–78	2%

Table 20.3 SCAN:3A Tests for Auditory Processing Disorders supplementary tests

	Raw score	Standard score	Confidence interval	Percentile rank
Competing Words–Free Recall (CW-FR)	29	9	6 – 12	37%
Auditory Figure-Ground (AFG) + 8	28	1	1 – 4	1%
Auditory Figure-Ground (AFG) + 12	36	6	2 – 10	1%
Time Compressed Sentences (TCS)	53	4	1 – 7	53%

The results in Table 20.4 indicate an LE advantage on six of the subtests. This finding is substantially abnormal, indicating that language is processed in areas of the brain other than the left temporal lobe and confirms a neurological basis for NM's auditory, language, reading, and learning problems.

The SSW findings are reported in Table 20.5, and the results are consistent with the results from the SCAN-3A test battery. All conditions of the SSW—Right Non-Competing (RNC); Right Competing (RC); Left Competing (LC); and Left Non-Competing (LCL)—are substantially poorer than the cutoff score for normal performance. In addition, the competing conditions of the SSW (RC and LC) reveal an LE advantage (i.e., less errors for LC and RC) as was also reported in several SCAN3-A tests.

Finally, the results for the DDT, reported in Table 20.6, are well below the cutoff score for normal performance. Again, these results are consistent with all other results. Unlike the other tests, however, the results of the DDT reveal an RE advantage of 10% When questioned about the DDT NM said, "I remember that by the time I had to repeat all of the numbers I forgot what I had heard and did a lot of guessing."

20.3 Questions to the Reader

1. What is NM's APD diagnosis? Are her findings consistent with a diagnosis of APD?
2. Is this a "rare form of auditory neuropathy" as diagnosed by a neurotologist, or something more?

Table 20.4 SCAN:3A Tests for Auditory Processing Disorders ear advantage (EA) summary showing the right ear advantage (REA) and left ear advantage (LEA) for each subtest with the cumulative percentage of the finding.

	Ear advantage typical (Y or N)			Typical (Y or N)	Cumulative percentage
Test	RE score − LE score		= EA		
Auditory Figure-Ground 0	13	8	5	N	5% REA
Competing Words–Free Recall	14	15	− 1	Y	
Filtered words	10	12	− 2	N	10% LEA
Competing Words–Directed Ear RE	8	11	− 3	N	2% LEA
Competing Words–competing ear LE	13	11	2	Y	
Competing Sentences	26	27	− 1	N	15% LEA
Auditory Figure–Ground + 8	14	14	0	Y	
Auditory Figure–Ground + 12	17	19	− 2	N	10% LEA
Time Compressed Sentences	26	27	− 1	N	10% LEA

Table 20.5 The SSW Test showing the number of errors for NM and the errors cutoff normative score

Conditions	Results (number of errors)	Normative data (number of errors)
Right Ear Non-Competing (RNC)	3	.04
Right Ear Competing (RC)	8	.07
Left Ear Competing (LC)	5	1.2
Left Ear Non-Competing (LNC)	5	.3
Total	21	1.61

Note that the findings indicate a LEA in the competing words condition.

Table 20.6 Results for the Dichotic Digits Free Recall Test reporting overall abnormally low performance and a resulting 10% REA

	Right ear	-	Left ear	Ear advantage
Percent correct	65%		55%	10% REA

3. What is the significance of the LE advantage revealed on several of the monaural degraded and dichotic speech tests?
4. Is this an "auditory specific" APD?
5. What habilitation indicator is indicated by the 8.5 dB SNR and low performance on the Auditory Figure-Ground (AFG) tests of SCAN-3A?
6. What implications does this case have for counseling of young patients with problems of reading, language, and audition?

20.4 Discussion of Questions to the Reader

1. What is NM's APD diagnosis? Are her findings consistent with a diagnosis of APD?
   Multiple findings of low performance on several tests of monaural degraded speech and dichotic speech measures indicate that NM has an abnormal neurological pathway that does not work efficiently for processing of auditory information.
2. Is this a "rare form of auditory neuropathy" as diagnosed by a neurotologist or something more?
   NM is more likely to have a complex set of neurological abnormalities from the auditory nerve to the auditory cortex and association areas of the brain. These findings are greater than a more "simple" diagnosis of auditory neuropathy, though NM displays those findings as well.
3. What is the significance of the LE advantage findings on several monaural degraded and dichotic speech tests?
   The finding of LE advantage on both monaural degraded and dichotic speech tasks indicate that auditory and language is not being processed in the typical left hemisphere. The finding of LE advantage on dichotic measures has been shown through functional magnetic resonance imaging (fMRI) studies to indicate processing of auditory information is occurring in areas of the brain that are not designed to manage these skills, which therefore works inefficiently and is to the affected person's educational disadvantage. Audiologists who fail to calculate ear advantage when performing tests of central auditory processing miss one of the more powerful clinical indicators of auditory pathway neurological abnormalities.
4. Is this an "auditory specific" APD?
   Clearly the problems of reading, language, and auditory processing are indicators of cross modality abnormalities that are not auditory specific. In fact, most of the individuals seen for auditory processing evaluation will have language and reading problems, and there are few documented cases of "pure" APD reported in the literature.
5. What habilitation indicator is indicated by the 8.5 dB SNR and low performance on the AFG tests of SCA-:3A?
   Abnormal performance on AFG tests provide evidence of the need for classroom management that may include reducing the distance of the primary signal and level of background noise, preferential seating, and recommendations for hearing assistive technology (HAT) devices in the classroom. When recommendations for HATs are made, care must be taken to educate not only the user but teachers who must be part of the team that makes this recommendation successful.
6. What implications does this case have for counseling of young patients with problems of reading, language, and audition?
   There are many situations where counseling of realistic expectation for academic success are indicated. Audiologists,

however, should be careful to avoid painting such a bleak picture of future success as to discourage parents, teachers, and especially the affected person that there is no hope for academic success and an intellectual life. To do so could work to discourage persons from trying and could have a long-term effect on their lives. Every effort should be made to identify positive sources of help to accommodate, compensate, and remediate the conditions that stand in the way of success.

20.5 Additional Discussion to the Reader

What can the reader learn from this case report? One of the important findings is the confirming evidence that intrauterine and early exposure to street drugs has lifelong consequences, including multiple comorbid problems of memory, attention, reading, speed of processing, language, and audition. The APD test battery confirmed many of the auditory complaints described in NM's educational case history. The entire battery provides clues to a generalized neurological pathway that does not work efficiently for listening, language, and reading function. Although NM did not receive an early diagnosis of her academic problems and had no intervention at any time, she was able to do well in school under certain strict classroom conditions. That is, a quiet, organized, and highly disciplined classroom structure worked to NM's advantage while she was in school. Beyond that, this case demonstrates that an intelligent and determined person can sometimes achieve a high level of success in spite of these sensory and cognitive factors. One important message for audiologists involved in evaluating APDs is that it is important to be judicious when counseling patients about possible future achievements in life in the face of significant sensory and cognitive factors.

20.6 Diagnosis and Recommended Treatment

Results of this APD test battery confirmed the presence of significant abnormalities in the functional performance of NM's auditory nervous system. Further, the findings were consistent with a neurological basis for her auditory processing, language, and reading problems. The presumed cause of her neurological problems was the intrauterine and early-life intake of street drugs due to her birth mother's substance abuse.

At this stage of life, NM has developed compensatory techniques that serve her well in her personal and professional life. She now has a doctoral degree and works with children with disabilities. It is unlikely that any specific remediation would be useful in changing her physiological status. The best option is for her to continue to use those strategies that work in her behalf, and to be aware of developing technology that will further assist her in the future.

20.7 Outcome

NM has earned a doctorate and a successful personal and professional life. She has used her intellectual gifts and determination to overcome multiple factors that might have prevented academic and personal success.

20.8 Acknowledgment

The author wishes to acknowledge Sally Vollner, a third-year AuD student at the University of Cincinnati, who obtained the audiometric results reported in this case report.

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21 Long-Term Effects of Cisplatin and Cranial Radiation Therapy on Hearing

Jennifer Listenberger

FJ is a 10-year male survivor of nasopharyngeal cancer. He has a history of chronic serous otitis media and progressively deteriorating hearing loss.

21.1 Clinical History and Description

FJ has been a patient for the past 10 years. FJ has been treated by otolaryngologists and is a hearing aid patient. He came to the audiology clinic for an audiological examination due to perceived bilateral hearing loss, which FJ said was due to chemotherapy.

Clinical notes from previous visits revealed FJ had experienced aural fullness and bleeding from the nose for several months in 2002. He noted ear drainage and had a pressure-equalization (PE) tube placed in the right tympanic membrane. A computed tomographic (CT) scan was completed, and a nasopharyngeal mass was found. FJ was diagnosed with squamous cell carcinoma of the nasopharynx in June 2002. Concurrent cisplatin and radiation therapy was recommended. The therapy plan commenced with one cycle of cisplatin and fluorouracil (5-FU), followed by concurrent cisplatin and radiation therapy with two cycles of cisplatin and 5-FU along with radiation of the head and neck administered from July through September 2002. Treatment ended with two cycles of postirradiation cisplatin and 5-FU with the final cycle administered in October 2002. Information regarding specific dosage of radiation and medication was not available.

FJ reported being provided with information regarding side-effects of the cisplatin but says he was not scheduled for an audiological examination at any time before, during, or after his treatment. FJ states there was no significant history of hearing loss prior to his diagnosis of nasopharyngeal cancer. FJ reported that he began to perceive tinnitus in his left ear, hearing loss, and difficulties communicating during his treatments. There was no history of noise exposure or pathology of the ear prior to the symptoms that arose as a result of his treatments associated with the cancer. Communication difficulties and perceived hearing loss progressed over the few months (June through October 2002) treatment was administered. FJ was fitted with bilateral behind-the-ear (BTE) amplification in 2003.

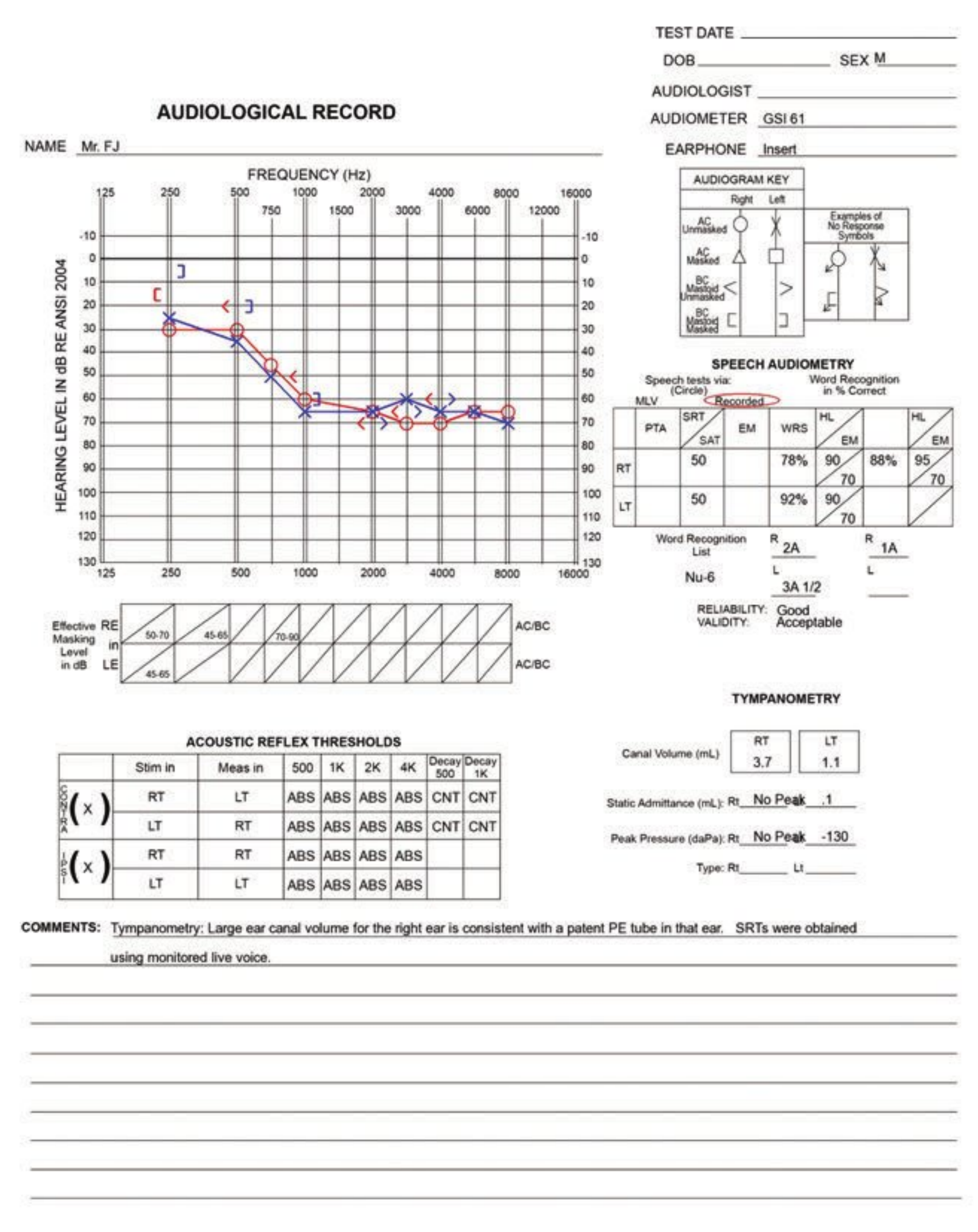
In addition to these problems, FJ has also had intermittent difficulties with otitis media with effusion. He has had myringotomy tubes inserted bilaterally several times. Chronic serous otitis media has persisted due to persistent eustachian tube dysfunction as a result of cranial radiation. FJ's hearing has progressively deteriorated and has also fluctuated as a result of middle ear involvement. He continues to use hearing aids bilaterally, and the hearing aids have been reprogrammed to accommodate changes in his hearing. FJ does well in quiet and uses a personal infrared hearing assistive technology (HAT) device when listening to his television. He is currently considering all options to improve communication.

21.2 Audiological Testing

Eighteen audiological examinations have been completed over the past 10 years. The initial audiological examination was completed in December 2002, which was 2 months following completion of treatment (Fig. 21.1). The results from this examination reveal a bilateral symmetric mild sloping to moderate sensorineural hearing loss (SNHL) through 750 Hz sloping to a moderately severe SNHL at 1000 through 8000 Hz. There is an air–bone gap present for the left ear at 250 to 500 Hz. Speech recognition thresholds (SRTs) reveal a moderate loss in the ability to receive speech bilaterally, and word recognition scores (WRSs) reveal slight difficulty for the right ear and normal ability in the left ear. The left ear presents with normal ear canal volume, but static admittance is hypocompliant and the peak pressure is negative, suggesting middle ear involvement. The right tympanogram has a flat configuration with a large ear canal volume, which is consistent with a patent PE tube. Acoustic reflex thresholds (ARTs) are absent bilaterally at 500 to 4000 Hz to contralateral and ipsilateral stimulation.

Intermittent eustachian tube dysfunction continued, and several audiological examinations were completed per physician referral and FJ's perceived symptoms. Over the repeated audiological examinations, bone conduction thresholds and WRSs remained stable for several years. Each time a decrease in hearing was perceived by FJ, it was a change in FJ's air conduction thresholds. The otitis media with effusion occurred bilaterally intermittently. When treatment for middle ear involvement was effective, FJ's air conduction thresholds would improve across all frequencies, and his hearing loss would resolve from a mixed hearing loss to SNHL as was reported in the initial audiological examination.

Fig. 21.2 reports on an audiological examination completed in June 2007, which was 5 years after the initial evaluation in 2002 and his initial diagnosis of nasopharyngeal cancer. Review of all of the previous audiological examinations confirms that this is the first audiogram revealing any significant decrease in bone conduction and air conduction thresholds at 3000 Hz and above without any known middle ear involvement. Unfortunately, there was a 3-year period between August 2004 and the audiogram reported in Fig. 21.2 where no audiological examinations were performed. This 3-year delay in follow-up audiological examinations made it difficult to determine when bone conduction thresholds began to decrease. FJ made appointments with his otologist for follow-up during the 3-year period but did not report any changes in his hearing. FJ was extremely ill and only scheduled office visits as needed. He made very few appointments regarding his amplification. All audiometric examinations between 2002 and 2004 revealed stable high-frequency bone conduction thresholds. At 2000 to 4000 Hz bone conduction thresholds were consistently between 60 and 70 dB HL. High-frequency air conduction thresholds were also stable when mixed hearing loss was not evident. As reported in Fig. 21.2, air conduction thresholds in



AUDIOLOGICAL RECORD

NAME Mr. FJ

TEST DATE
DOB
SEX M
AUDIOLOGIST
AUDIOMETER GSI 61
EARPHONE Insert

Effective Masking Level in dB: RE in 50-70, 45-65, 70-90 AC/BC; LE in 45-65 AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		50		78%	90 / 70	88%	95 / 70
LT		50		92%	90 / 70		

Word Recognition List Nu-6: R 2A, L 3A 1/2; R 1A, L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	3.7	1.1

Static Admittance (mL): Rt No Peak Lt .1
Peak Pressure (daPa): Rt No Peak Lt -130
Type: Rt Lt

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS: Tympanometry: Large ear canal volume for the right ear is consistent with a patent PE tube in that ear. SRTs were obtained using monitored live voice.

Fig. 21.1 Initial audiological examination, completed 4 months after FJ's first dose of cisplatin and 2 months after his last dose of cisplatin.

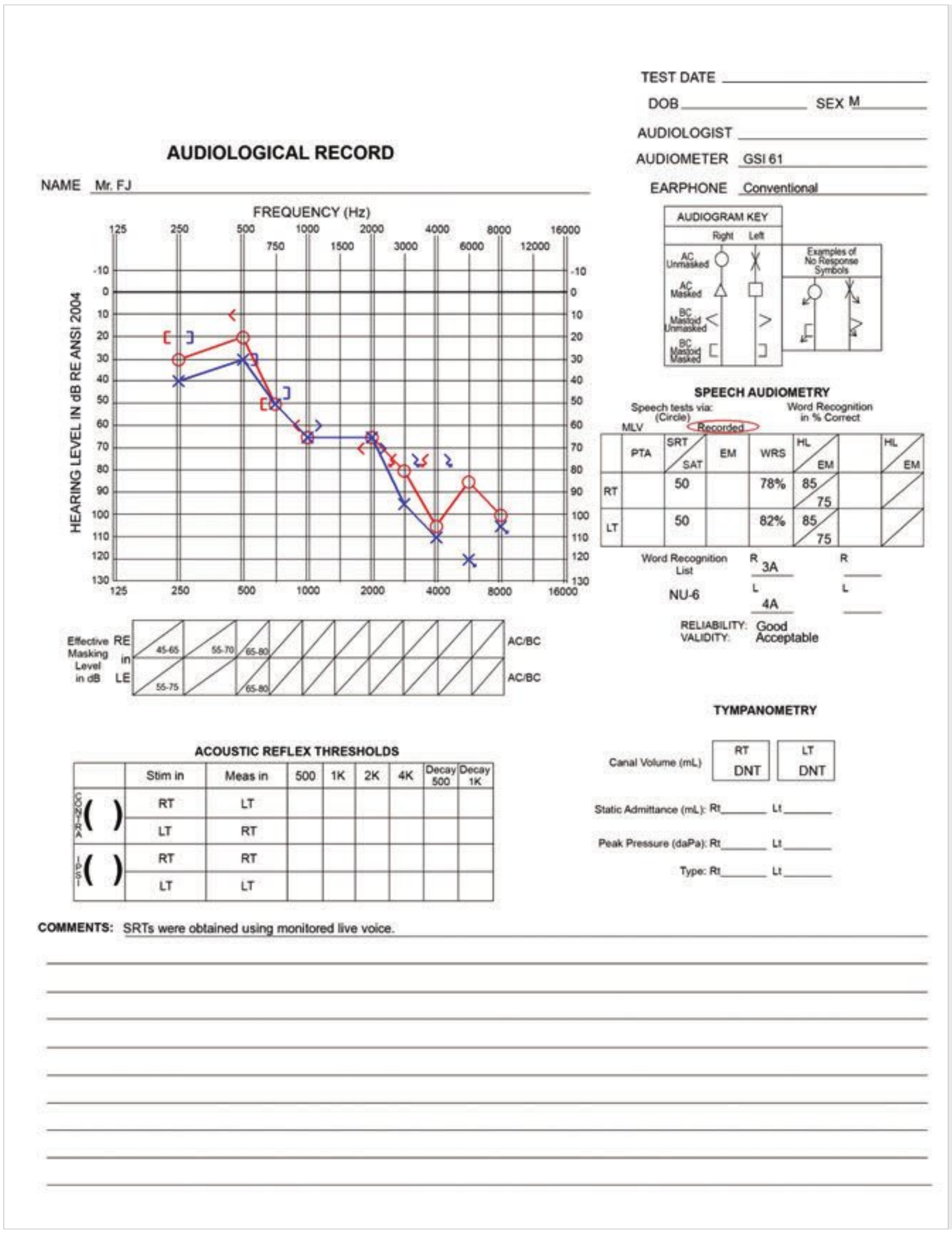
the left ear at 3000 to 8000 Hz decreased from moderately severe at the initial evaluation to profound with no measurable response at 6000 to 8000 Hz. Air conduction thresholds in the right ear decreased at 3000 to 8000 Hz from moderately severe to severe and then sloping to a profound SNHL. No response is measured for bone conduction thresholds at 3000 to 4000 Hz. WRSs revealed slight difficulty in the ability to recognize speech bilaterally.

Audiological examinations were completed annually from 2007 to the present due to FJ's perceived increased hearing difficulties and continuous bouts with chronic serous otitis media. He was also having increased difficulty with communication and felt he was receiving less benefit from his hearing aids. The most recent audiological examination was completed in December 2012 (Fig. 21.3). As can be seen in Fig. 21.3 there has been a significant decrease bilaterally in WRSs, which have decreased from slight difficulty to very poor ability to recognize speech. The greatest change in WRSs occurred over the past 2 years. FJ currently has a severe to profound mixed hearing loss in the right ear and a profound mixed hearing loss in the left ear. Tympanometry could not be completed due to the inability to maintain an appropriate seal.

In summary, over a 10-year period, there has been a gradual decrease in high-frequency thresholds and, more recently, a rapid decrease in low- and midfrequency hearing thresholds. There is no measurable response at 4000 to 8000 Hz bilaterally. Bone conduction thresholds at 250 to 1000 Hz remained relatively stable, but there is no measurable bone conduction threshold at 2000 to 4000 Hz.

21.3 Questions to the Reader

1. Given that treatment with a known ototoxic medication was recommended, when should the initial audiometric examination have been completed?
2. If an ototoxicity monitoring protocol had been followed, what differences in hearing thresholds and treatment(s) might have occurred?
3. Does head and neck radiation add to the importance of conventional and long-term audiological monitoring?
4. Based on the most current audiological examination what further recommendations can be made for FJ to improve communication?



AUDIOLOGICAL RECORD

NAME Mr. FJ

TEST DATE

DOB

SEX M

AUDIOLOGIST

AUDIOMETER GSI 61

EARPHONE Conventional

Effective Masking Level in dB				
RE in	45-65	55-70	65-80	AC/BC
LE	55-75		65-80	AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		50		78%	85 / 75		
LT		50		82%	85 / 75		

Word Recognition List NU-6 R 3A L 4A R L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	DNT	DNT

Static Admittance (mL): Rt______ Lt______

Peak Pressure (daPa): Rt______ Lt______

Type: Rt______ Lt______

COMMENTS: SRTs were obtained using monitored live voice.

Fig. 21.2 Audiological evaluation completed in 2007, which was 5 years post–cancer diagnosis and completion of treatment. This was the first audiological examination to report changes in bone conduction thresholds.

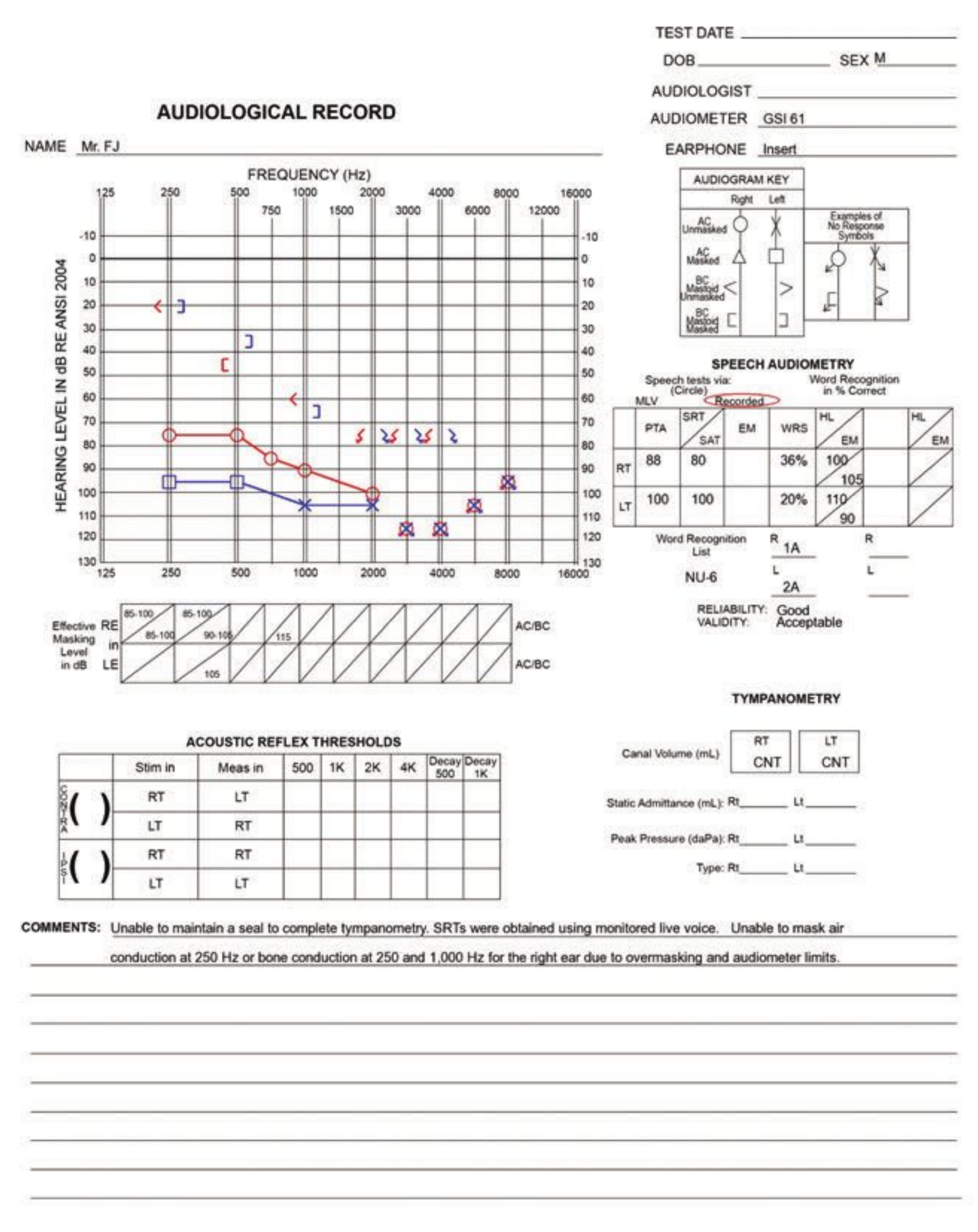
21.4 Discussion of Questions to the Reader

1. Given that treatment with a known ototoxic medication was recommended, when should the initial audiometric examination have been completed?

   Ideally, FJ's initial audiological examination should have been completed prior to the initiation of any ototoxic treatment in order to establish a baseline. According to American Speech-Language-Hearing Association (ASHA) guidelines, the baseline test for a patient receiving ototoxic medication should be completed prior to treatment or no later than 24 hours after treatment has begun.

   Cisplatin is frequently used for treatment of head and neck cancer and is one of the most ototoxic clinical compounds. The deleterious effect of cisplatin on the auditory and vestibular systems is well documented. Also, the importance of implementing an ototoxic monitoring protocol is equally well documented. A monitoring protocol suggested for a patient receiving cisplatin is that hearing be measured within 24 hours preceding each cycle of cisplatin, immediately after the final cycle, and at 3 and 6 months post-treatment. The frequency of ototoxic monitoring performed during treatment will depend on the patient's specific drug regimen and perceived changes in hearing. According to FJ, there were no recommendations for audiological examination before or during treatments. Unfortunately, in FJ's case an audiological examination was not obtained until 4 months after initiation of treatment and 2 months after treatment was completed when FJ requested an audiometric examination.
2. If an ototoxicity monitoring protocol had been followed, what differences in hearing thresholds and treatment might have occurred?

   The primary purpose of an ototoxic monitoring protocol for patients receiving ototoxic medication is for early identification of changes to the auditory or vestibular systems before damage occurs. Symptoms (i.e., decreased hearing and/or dizziness) can present immediately after the first treatment with an ototoxic medication or symptoms can be delayed for several weeks or months. As discussed earlier, a baseline audiological examination should be completed before initiation of treatment with an ototoxic agent, and this is followed



TEST DATE

DOB SEX M

AUDIOLOGIST

AUDIOMETER GSI 61

EARPHONE Insert

AUDIOLOGICAL RECORD

NAME Mr. FJ

Effective Masking Level in dB					
RE in	85-100 / 85-100	85-100 / 90-105		115	AC/BC
LE		105			AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	88	80		36%	100 / 105		
LT	100	100		20%	110 / 90		

Word Recognition List NU-6 R 1A L 2A R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	CNT	CNT

Static Admittance (mL): Rt Lt

Peak Pressure (daPa): Rt Lt

Type: Rt Lt

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: Unable to maintain a seal to complete tympanometry. SRTs were obtained using monitored live voice. Unable to mask air conduction at 250 Hz or bone conduction at 250 and 1,000 Hz for the right ear due to overmasking and audiometer limits.

Fig. 21.3 Final audiological examination, which was completed 10 years after his initial audiological examination.

by a hearing monitoring protocol to monitor hearing as the patient undergoes treatment and for several months after treatment is completed. Ototoxic therapies typically reveal initial damage in the basal region, or high-frequency region, of the cochlea. Ideally, an ototoxic monitoring protocol will include high-frequency audiometry to provide information for early detection. In order for an ototoxicity monitoring program to be successful there must be communication between the audiologist and the medical professionals prescribing the ototoxic therapies.

It is difficult to speculate what change in the dosage or in the drug therapy, if any, would have been recommended for FJ had ototoxic monitoring been followed. At the very least, a baseline audiological examination should have been ordered and completed. Also, FJ's audiologist would have been involved in counseling FJ about the effects of the ototoxic medication and the importance of reporting any changes in his hearing or balance to his oncologist. If a monitoring protocol is in place and hearing loss is detected then it is the audiologist's responsibility to inform the treating physician of these changes. This information will provide the physician the opportunity to decide whether or not the treatment regimen should or could be adjusted by changing the therapeutic agent or altering the dose. If the treatment plan cannot be altered then the audiologist is responsible for monitoring the hearing of the patient, counseling the patient on the effects of the ototoxic agent, and providing rehabilitation in the form of communication strategies, HAT, or hearing aids as deemed necessary. From an audiological and communication viewpoint, an audiologist could have introduced and recommended assistive technology for FJ to use at home or during office visits if the patient was not yet using amplification. The audiologist and physician could have counseled FJ on the potentially progressive nature of his hearing loss due to the use of cisplatin and provided realistic expectations and communication strategies for the patient and his family. The patient must be educated on the potential hearing loss resulting from the prescribed ototoxic medication and encouraged to report all changes in hearing or balance.

3. Does head and neck radiation add to the importance of conventional and long-term audiological monitoring? Radiation can affect all structures of the auditory system, including the external ear through the central auditory pathways. The damage incurred can be dependent upon the site

of lesion being irradiated and the dose of radiation. In the case of nasopharyngeal cancer, the inner, outer, and middle ear can all be in the field of radiation. Due to the site of FJ's tumor and its proximity to the ear, the cochlea and eustachian tube can receive the same dose of radiation as the tumor or possibly more. The tumor itself can invade the middle ear and cause symptoms as was the case with FJ and his early symptoms of aural fullness. Swelling of the mucosal lining and blockage of the cartilaginous portion of the pharyngeal opening of the eustachian tube can be a product of radiation potentially resulting in chronic otitis media. The SNHL associated with cranial radiation can appear several months or even several years postirradiation. The SNHL is usually poorer in the higher frequencies and can be progressive in nature. The mechanism of the SNHL due to cranial irradiation is not completely clear, but several theories propose the hearing loss is due to compromised blood supply to the cochlea causing damage to the hair cells.

Currently there are no published standards or guidelines for monitoring ototoxicity due to head and neck radiation, but there have been numerous studies that have investigated hearing loss due to head and neck radiation. It is well documented that cranial radiation alone can negatively impact the auditory system, and ototoxic monitoring should be implemented during this treatment as well. The combination of the ototoxic chemotherapy and cranial radiation should have warranted the initiation of a monitoring protocol for FJ. A baseline audiological examination should have been completed prior to any treatment, but, in the case of FJ, his initial examination should have been completed before his first round of cisplatin and then just before his first dose of radiation. The studies that have reported SNHL as a result of radiation stated that hearing loss can occur during or immediately after radiation therapy or even up to 5 to 8 years after radiation therapy.

4. Based on the most current audiological examination what further recommendations can be made for FJ to improve communication?
   FJ is experiencing great difficulty with communication. He uses his hearing aids but has great difficulty correctly recognizing speech even in quiet listening situations. FJ is considering personal FM technology to use with his hearing aids. The severity of the pure-tone thresholds, the poor WRSs, and the perceived lack of benefit from traditional amplification justify the recommendation for an evaluation for a cochlear implant (CI). FJ did undergo a CI evaluation and was found to be a good candidate. He has many health issues and is concerned about surgery and as a result he decided not to pursue the CI. FJ has planned to continue using his hearing aids, and he will follow-up with his audiologist when he is interested in pursuing FM technology.

21.5 Outcome

As discussed earlier, chronic serous otitis media persists, and FJ continues to experience great difficulty with communication due to the severity of his hearing loss. FJ continues follow-up visits with his otologist regarding the persistent conductive component to his hearing loss and possible middle ear effusion. FJ recently had a CI evaluation, and it was recommended that he continue the preoperative process. Annual evaluations have revealed no recurrence of the nasopharyngeal cancer; however, FJ remains quite ill. He has concerns regarding surgery and his greatest concern is the anesthesia. FJ again decided against pursuing the CI due to his health and he continues using his hearing aids while considering a personal FM system.

21.6 Key Points

1. According to the electronic medical records and FJ's case history, no audiologist was involved when FJ was initially diagnosed with nasopharyngeal cancer and treatment with an ototoxic medication was planned. The risk of hearing loss due to cisplatin therapy is well documented, and audiologists need to be included in the care and treatment of patients receiving ototoxic medications.
2. Currently, there are no national guidelines for audiological monitoring protocols for monitoring the hearing of patients receiving head and neck radiation. Research does indicate that cranial radiation alone can cause hearing loss, and hearing loss can occur immediately after the start of treatment or even as long as 5 years after treatment is completed. It is important to understand the medical treatments patients have undergone and continue to receive. It is very difficult for audiologists to separate the ototoxicity caused by cisplatin and the ototoxicity caused by radiation or determine which agent was the most damaging to FJ's hearing. The most important message is to understand and combat the long-term effect and the need for continued follow-up and monitoring even several years after treatment with these two ototoxic agents.

Suggested Reading

American Speech-Language-Hearing Association (ASHA). Guidelines for the audiologic management of individuals receiving cochleotoxic drug therapy. 1994. http://www.asha.org/policy/GL1994–00003.htm

Bass J, White SAT. Tutorial: Radiation-Induced Hearing Loss. Audiology Today 2008; 20: 22–24http://www.audiology.org/resources/audiologytoday/Documents/AudiologyToday/2008ATMarApr.pdf

Butler SK. Cisplatin: forty-five years later and what we still do not know about ototoxicity. Semin Hear 2011; 32: 229–235https://www.thieme-connect.de/ejournals/pdf/10.1055/s-0031-1286617.pdf?update=true

Fausti S, Helt W, Gordon J, Reavis K, Phillips D, Konrad-Martin D. Audiologic monitoring for ototoxicity and patient management. In: Campbell K, ed. Pharmacology and Ototoxicity for Audiologists. Clifton Park, NY: Thomson Delmar Learning; 2007:230–251

Jereczek-Fossa BA, Zarowski A, Milani F, Orecchia R. Radiotherapy-induced ear toxicity. Cancer Treat Rev 2003; 29: 417–430

Rybak L. Cancer and ototoxicity of chemotherapeutics: audiologic monitoring for ototoxicity and patient management. In: Campbell K, ed. Pharmacology and Ototoxicity for Audiologists. Clifton Park, NY: Thomson Delmar Learning; 2007:139–162

22 One Patient with Two Audiological Examinations and Two Different Outcomes

Jennifer Listenberger

LA had two audiological examinations within 1 year from two different clinics and the results from the two examinations differed significantly, resulting in distinctly different sets of recommendations.

22.1 Clinical History and Description

LA is a 27-year-old female who presents with a long-standing history of hearing loss in her left ear and what she refers to as “sensitive hearing.” LA reports that many external sounds such as chewing, people clearing their throats, certain voices, noise in crowds, and coins banging together, bother her. She says her tolerance for these sounds is very low, and sometimes the sounds are painful. She has difficulty recognizing speech in background noise and will position people on her right side so she can hear better. Reportedly, her misophonia (i.e., abnormally strong negative reaction to specific soft sounds) and hearing loss have been present since childhood.

LA reports no significant history of ear pathology. As far as LA knows, all birth, delivery, and developmental milestones were within normal limits. There is some familial hearing loss. Her brother reportedly has hearing loss, but there are no other significant issues related to family history. LA reports a history of taking medication for treatment of anxiety and obsessive compulsive disorder. LA has been taking this medication for several years and recently ceased taking the medication because she is starting a family. There is a recent onset of dizziness and increase in the misophonia, but she feels these are related to discontinuing the antianxiety medication. LA reports that the external sounds and background noise do not cause dizziness. LA does not use any hearing protection when she is out in public because she can tolerate the sounds without earplugs.

LA scheduled an appointment at the current clinic to obtain a second opinion regarding her hearing loss and “hearing sensitivity.” LA had an audiological examination approximately 1 year before scheduling the appointment in the current audiology clinic. The report from the previous clinic stated that LA had an asymmetric sensorineural hearing loss with mild low-frequency sensorineural hearing loss through 2000 Hz rising to within normal limits in the right ear and a mild sensorineural hearing loss in the left ear that is flat in configuration. Tympanometry was reported to be “peaked,” although no additional specific information was provided. The recommendation by the audiologist at the previous clinic was for LA to return to her referring physician.

According to LA, she was not surprised by the diagnosis of hearing loss due to her perception of communication difficulties since childhood. LA reported that she felt she will need a hearing aid at some point. There are no other audiological examinations available for comparison, and LA says she has not had her hearing evaluated since she was a teenager.

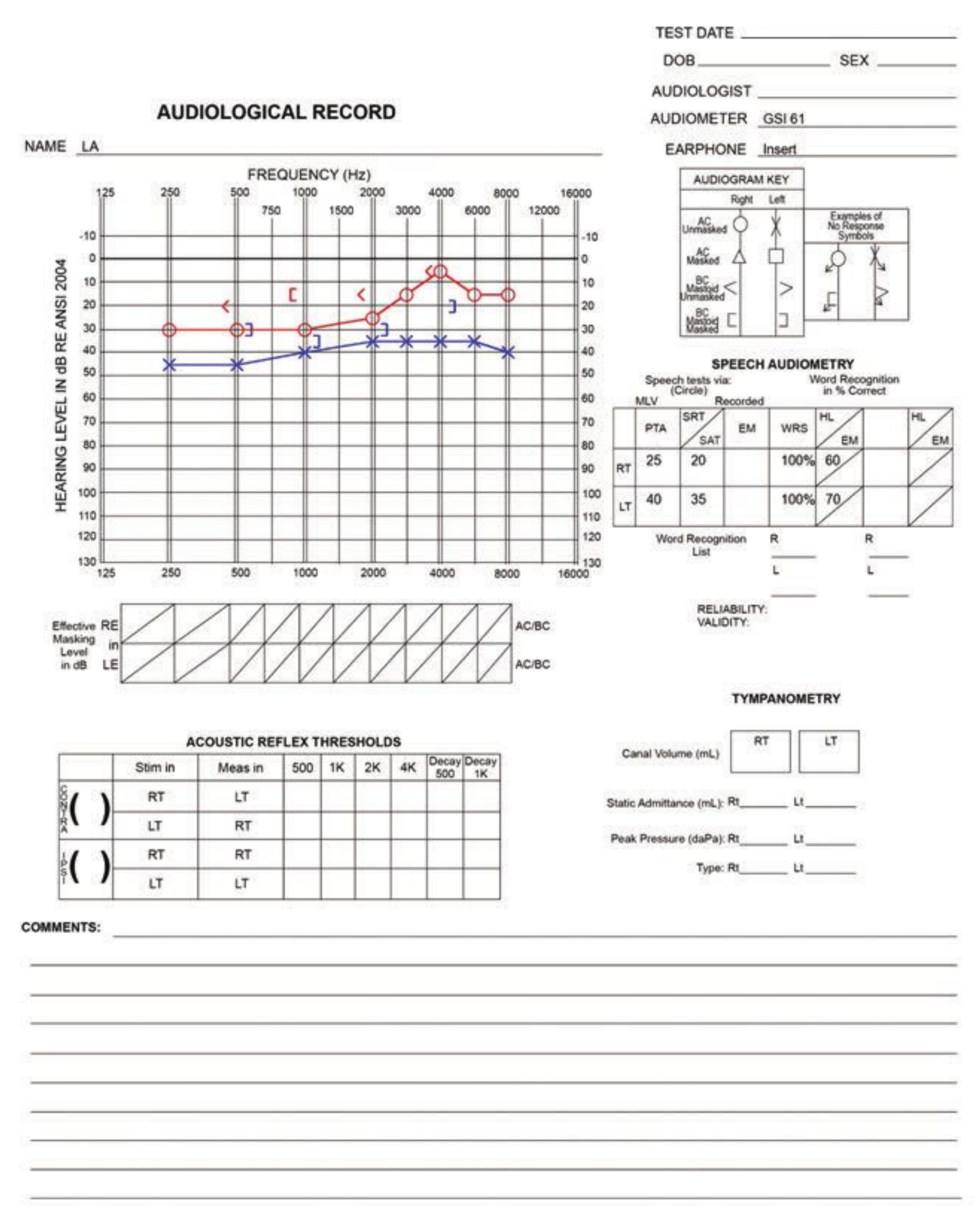
22.2 Audiological Testing

Fig. 22.1 reports the results of LA’s initial audiological examination from the other clinic. This audiological examination reveals a moderate rising to a mild mixed hearing loss in the left ear and a mild low frequency sensorineural hearing loss in the right ear rising to within normal limits at 3000 to 8000 Hz. Speech recognition thresholds (SRTs) revealed a slight loss in the ability to receive speech in the right ear and a mild loss in the ability to receive speech in the left ear, and both are in agreement with the pure-tone averages (PTAs). Word recognition scores (WRSs) indicated normal ability to recognize speech bilaterally. The audiogram reveals a mixed hearing loss in the left ear, but the report accompanying the audiogram from the clinic where the initial audiogram was completed interpreted the results as sensorineural. Immittance audiometry that would include tympanometry, acoustic reflex thresholds (ARTs), and acoustic reflex decay were not reported.

The results from the audiological examination completed at the current clinic are reported in Fig. 22.2. This audiogram reveals normal hearing in the right ear with a slight notch at 500 Hz. Results for the left ear reveal a mild conductive hearing loss with a mixed component at 2000 Hz (i.e., Carhart notch). SRTs reveal a slight loss in the ability to receive speech in the right ear and a mild loss in the ability to receive speech in the left ear and are in agreement with the PTAs bilaterally. WRSs reveal normal ability to recognize speech bilaterally.

Immittance audiometry was completed with tympanometry revealing normal tympanic membrane mobility bilaterally. ARTs were absent bilaterally to ipsilateral and contralateral stimulation at 500 to 4000 Hz. Absent ARTs to the right ipsilateral ART stimulus is not consistent with the normal hearing in that ear. Absent ARTs to the left ipsilateral stimulus and the absent ARTs to the right and left contralateral stimulation were consistent with the conductive hearing loss in the left ear.

In summary, the initial audiological examination at the other clinic revealed an asymmetric hearing loss with a mixed hearing loss in the left ear and a sensorineural hearing loss in the right ear. The more current audiological examination revealed normal hearing in the right ear and a mild conductive hearing loss in the left ear. The unmasked bone conduction thresholds and left air conduction thresholds remained unchanged between the two examinations. There were significant improvements in the left bone conduction thresholds at 500, 1000, 2000, and 4000 Hz and in the right air conduction thresholds at 250 to 2000 Hz in the current examination compared with the initial examination. Masked left bone conduction thresholds improved in the current examination by 20 and 25 dB at 500 and 1000 Hz, respectively, and 10 dB at 2000 and 4000 Hz in the current examination compared with the initial examination, and the air conduction thresholds for the right ear improved by 10 to 15 dB at 250 to 2000 Hz, respectively.



TEST DATE ______
DOB ______ SEX ______
AUDIOLOGIST ______
AUDIOMETER GSI 61
EARPHONE Insert

AUDIOLOGICAL RECORD

NAME LA

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	25	20		100%	60		
LT	40	35		100%	70		

Word Recognition List R ______ L ______ R ______ L ______

RELIABILITY:
VALIDITY:

TYMPANOMETRY

Canal Volume (mL) RT ______ LT ______
Static Admittance (mL): Rt______ Lt______
Peak Pressure (daPa): Rt______ Lt______
Type: Rt______ Lt______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: ______

Fig. 22.1 Initial audiological examination completed at a different audiology clinic.

22.3 Questions to the Reader

1. The two audiological examinations differ in the information recorded on the report form. What information, if any, is missing from the initial audiological examination?
2. What audiological tests should have been completed at the initial examination to provide a more comprehensive assessment of LA's hearing loss?
3. Because there is an asymmetry in the initial examination, what audiological test could have been completed to assist in confirming the presence or absence of a sensorineural or conductive hearing loss?
4. The amount of information reported between the two clinical sites is quite different. What factors could help explain the differences between the two audiological reports?

22.4 Discussion of Questions to the Reader

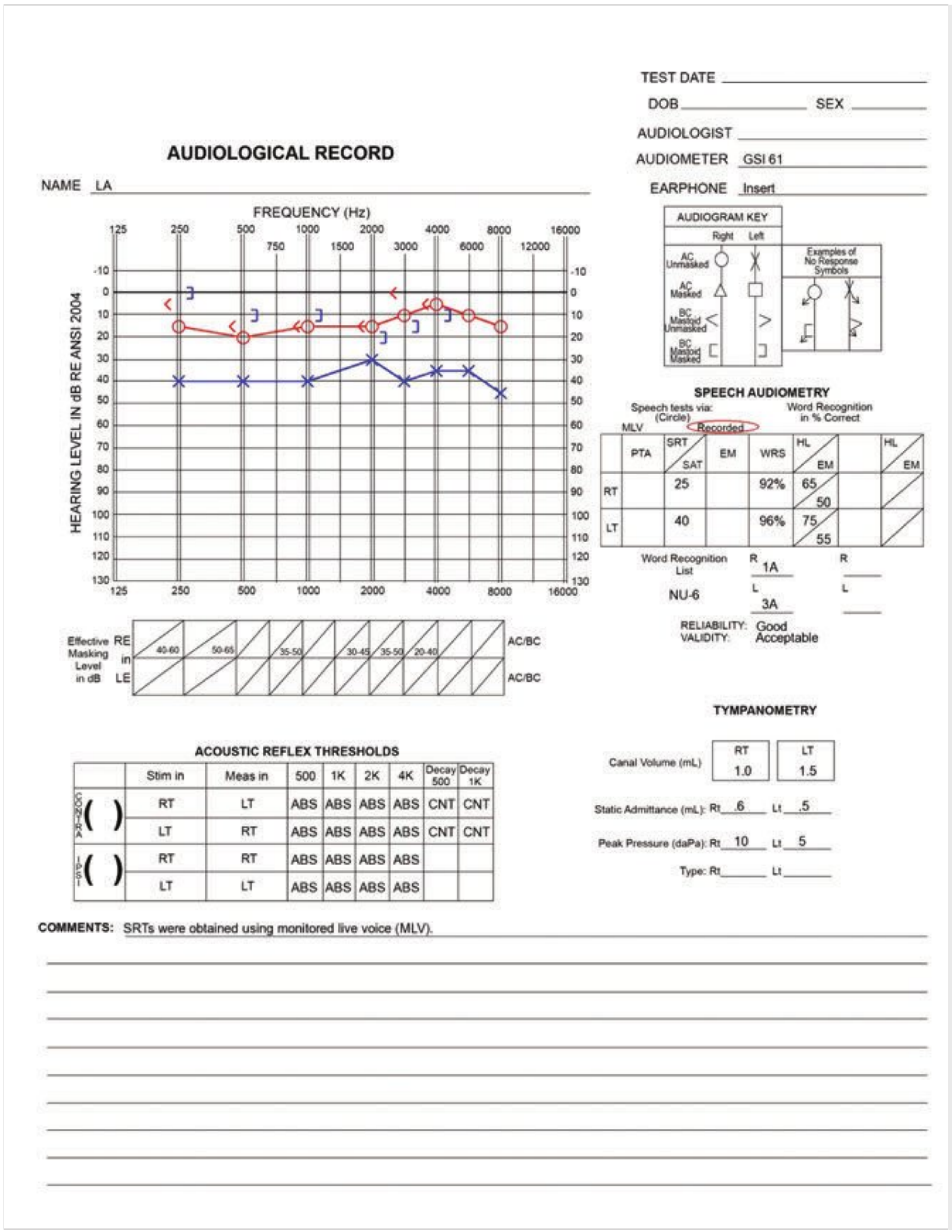
1. The two audiological examinations differ in the information recorded on the report form. What information, if any, is missing from the initial audiological examination?
   The presentation of the audiogram, including the recording of the results and the "appearance" of the audiometric symbols, is typically the first impression an audiologist makes when viewing the results obtained by another audiologist. The "neatness" of the audiogram and the documenting of all test results are essential. Written reports do not always accompany audiograms, and it is helpful to have as much important information as feasible recorded on the audiogram report form. Audiograms are often repeated for a number of reasons, and it is important for another audiologist to view any audiogram and be able to make accurate comparisons.

The initial audiogram (▯ Fig. 22.1) lacks essential information. The audiogram report form does not contain any information regarding the type of headphones/transducers used during the initial examination nor is there any documentation of patient response reliability or validity. Masking was used during bone conduction testing, and the masking levels were not noted. The audiologist did not document any essential information regarding the manner in which speech audiometry was performed. Also, the spaces to document the results from tympanometry, ARTs, and reflex decay test are blank in ▯ Fig. 22.1. There was no documentation to indicate whether these tests were attempted or completed.

When the type of transducer is not documented on the audiogram report form it is typically assumed that supra-aural headphones were used. The second audiogram report form (▯ Fig. 22.2) clearly states that insert earphones were used, although the specific ear tip style and size were not noted. All earphone transducers used with an audiometer are assumed to be calibrated to that audiometer; it should not make a difference, therefore, whether insert earphones or conventional supra-aural headphones were used. It is best practice, however, to document the transducer used if the results from several audiological examinations need to be compared.

Also, the initial audiogram report form (▯ Fig. 22.1) provides no information regarding which word test or word list was used or the presentation level of the stimuli for word recognition testing. It was also not reported whether a full or half-list was used. There was no indication of whether SRTs or WRSs were obtained using monitored live voice (MLV) or recorded speech. Many audiologists will, unfortunately, use MLV presentation to assess word recognition abilities. It is important to know what stimulus was used and how the stimulus was presented in order to appropriately compare test–retest data between audiologists and clinics.

It is important to remember that the results reported in ▯ Fig. 22.1 are assumed to be the initial audiological examination with no previous audiological results available for comparison. It is assumed that a comprehensive diagnostic examination was administered, including performance of immittance audiometry. The report from the initial clinic that accompanied the audiogram report form stated that tympanometry was completed and the tympanogram was "peaked." Reporting values of



TEST DATE

DOB SEX

AUDIOLOGIST

AUDIOMETER GSI 61

EARPHONE Insert

AUDIOLOGICAL RECORD

NAME LA

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT		25		92%	65/50		
LT		40		96%	75/55		

Word Recognition List NU-6 R 1A L 3A R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.0	1.5

Static Admittance (mL): Rt .6 Lt .5

Peak Pressure (daPa): Rt 10 Lt 5

Type: Rt Lt

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI ( )	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS: SRTs were obtained using monitored live voice (MLV).

Fig. 22.2 Audiological examination completed at our audiology clinic approximately 1 year after the initial audiological examination.

static compliance (mL), middle ear pressure (daPa), and ear canal volume (mL) and reporting classifications such as Type A, As, Ad, B, or C will provide information regarding the status of the middle ear. "Peaked" could indicate normal mobility, or a hypermobile eardrum, but this term could also imply a "peak" at negative pressure. There is no documentation in the space available for ART results. This implies that acoustic reflex testing was not completed. Any information provided on an audiogram regarding why a test was not completed or could not be completed is very important.

The masking levels for bone conduction testing on the initial audiogram were not reported. Both audiograms are variations of the format recommended by the American Speech-Language-Hearing Association (ASHA). This format has a space below the audiogram grid designated for reporting masking levels used in the nontest ear. It is not uncommon to those values omitted even when masking was used. Some may argue that audiologists should know how to mask effectively and that this information need not be recorded. Recorded masking levels are useful for teaching, follow-up testing, determining if appropriate masking levels were used, and making comparisons when reviewing test results.

2. What audiological tests should have been completed at the initial examination to provide a more comprehensive assessment of LA's hearing loss?
The results reported in Fig. 22.1 would be considered a comprehensive examination by some audiologists and otolaryngologists. It is the viewpoint of the author that the results reported in Fig. 22.1 do not represent an appropriate comprehensive initial audiological examination. It is the audiologist's responsibility to evaluate the patient's auditory system as completely as possible in order to make appropriate and accurate recommendations and referrals. ASHA's Preferred Practice Patterns for the Profession of Audiology state that a conventional audiological examination is used to "assess, evaluate, and monitor the status and function of the peripheral auditory system, which includes the external, middle, and inner ears as well as the auditory nerve." The lists of tests provided in this document include immittance audiometry as part of the clinical process for a comprehensive audiological examination.

   Immittance audiometry with ARTs was not reported for the audiological examination shown in Fig. 22.1. One purpose of immittance audiometry is to provide information regarding the status of the middle ear. Immittance audiometry may also be helpful for differential diagnosis regarding cochlear versus retrocochlear disease, and it is helpful in validation of the pure-tone audiogram. Tympanometry was completed as part of the initial audiological examination, but classification and specific values for static admittance (mL), ear canal volume (mL), and middle ear pressure (daPa) were not documented. Reporting of such results would have provided some information regarding the status of the middle ear.

   ART testing would have assisted the audiologist in the initial diagnostic evaluation. The original audiological report that accompanied the audiogram form described the loss as sensorineural. The hearing loss presented from the original audiogram report form in Fig. 22.1, however, is mixed as opposed to sensorineural in type of hearing loss. ART testing should have been completed to further evaluate the status of the middle ear system to confirm the conductive component. In this case, the audiologist considered the hearing loss to be sensorineural as opposed to mixed. An asymmetry was present and ARTs should have been completed to assist with differential diagnosis regarding cochlear or retrocochlear involvement.

3. Because there is an asymmetry in the initial examination, what audiological test could have been completed to assist in confirming the presence or absence of a sensorineural or conductive hearing loss?
   Physicians often use the Weber tuning fork test to help assess and/or confirm whether or not an asymmetric hearing loss is conductive or sensorineural. The Weber test is based on the theory that the stimulating tone will lateralize to the ear with the conductive component because the energy at that cochlea is greater or because there is a phase lead to that ear. When an asymmetry is present, an audiologist could perform a Weber test with the bone oscillator to assist with diagnosis. In LA's case, the otologist performed a tuning fork Weber test with a 512 Hz tone, and LA noted lateralization to the left. This finding assisted in the resolution of two conflicting test results. The Weber test is more reliable at frequencies below 1000 Hz than at higher frequencies. In this case report, there is an air–bone gap at 500 Hz in Fig. 22.1. If the hearing loss was sensorineural, the patient would have perceived tone lateralization to the right side. If a Weber test with the bone oscillator was completed and LA lateralized to the left side, then it might have alerted the audiologist to retest bone conduction thresholds or complete ART testing to assist with confirming the test results.

4. The amount of information reported between the two clinical sites is quite different. What factors could help explain the differences between the two audiological reports?
There were significant differences between the right air conduction thresholds and the left masked bone conduction thresholds between the initial and current audiological examinations. It is feasible that LA's hearing did not change over the course of a year, and the differences between the two audiological examinations could be a result of clinician error. Two factors could potentially explain the differences in the air and bone conduction thresholds between the two examinations.

   Air conduction thresholds were measured for both examinations using ER-3A insert earphones (Etymotic Research, Inc., Elk Grove Village, IL). A report accompanying the initial audiogram stated that insert earphones were used even though "transducer type" was not recorded on the audiogram report form. There are two types of ear tips that can be used with the insert earphone transducers: foam E-A-R ear tips and probe ear tips used for immittance audiometry that can be attached to the ER-3A inserts via an adapter. The foam ear tips can be inserted deeply into the ear canal and expand to help seal the sound pathway as delivered into the ear canal. The probe ear tips may not be inserted as deeply as the foam, but a seal should be attainable due to multiple sizes of tips available to accommodate almost any ear. Foam ear tips were used for the test completed in Fig. 22.2. Regardless of ear tip used, transducer placement within the ear canal is very important. If an insert tip is used and there is improper depth or an improper seal, then low-frequency energy could be lost or could leak out. This results in a need for a greater input level to elicit a threshold response resulting in falsely elevated air conduction thresholds.

There are no masking values reported on the initial audiogram. There is a possibility that overmasking occurred during bone conduction testing. Because masking values were not reported, it is difficult to determine if masking levels might be a cause for the differences between audiological results at the two examinations. The right air conduction thresholds are 10 to 15 dB better in Fig. 22.2 than in the initial audiogram (Fig. 22.1). The poorer air conduction thresholds reported in the initial audiogram could contribute to a higher starting point for masking levels before a masking plateau was established. When hearing loss is sensorineural, a false improvement in bone conduction thresholds in the low frequencies can occur when a headphone is coupled to the nontest ear in preparation for masking. This improvement in bone conduction thresholds is called the occlusion effect (OE). Additional levels of masking noise are typically added to initial masking levels to compensate for this additional energy at the cochlea. The occlusion effect can be measured individually or mean population values can be used. The mean values of 15 dB at 250 Hz, 15 dB at 500 Hz, and 10 dB at 1000 Hz are often used. In this case, if additional masking was applied to the right ear to compensate for a predicted occlusion effect, the masking level would be even higher. This could potentially cause the masking noise to cross and reach the cochlea of the test ear (the left ear in this case). This would falsely shift or elevate the bone conduction thresholds of the test ear.

22.5 Diagnosis and Recommended Treatment

LA originally scheduled an appointment with an otologist in the current clinic for a second opinion, but her initial audiological results were not available at the time of the appointment. The otologist referred LA for an audiological examination. After the audiological examination was completed a diagnosis of ossicular chain fixation was made for the left ear with an emphasis on the possibility of otosclerosis. The report from the otologist stated that the results from the measured ARTs helped to clarify the diagnosis. The fact that the Weber test lateralized to the left side, the left ipsilateral and contralateral ARTs were absent, air–bone gaps were present, and a Carhart notch was present at 2000 Hz helped the otologist arrive at the diagnosis that the ossicular chain was involved.

22.6 Outcome

The audiological examinations completed for this patient at the two sites were significantly different. When LA completed the initial examination, she was left with the recommendation that there was no need for medical or surgical intervention and amplification was the only treatment option due to the sensorineural hearing loss. The second audiological examination resulted in an entirely different outcome. The second audiological examination revealed a conductive hearing loss in the left ear and LA was provided with a surgical option to improve her hearing.

LA did not return to the clinic following her audiological examination and her second visit with the otologist. Unfortunately, at the time of the second visit, LA was pregnant and the otologist recommended that she return in 1 year for a follow-up examination and possible discussion regarding corrective surgery. There are no appointments scheduled for the near future.

22.7 Key Points

1. It is imperative for audiologists to remember that audiologists are diagnosticians who become investigators when administering a hearing examination. Audiologists are not simply quantifying hearing loss, but using the behavioral audiometric examination and immittance measures to investigate the auditory system as thoroughly as possible. These findings help clinicians decide what further recommendations are needed regarding electrophysiological testing, physician referrals, amplification, aural rehabilitation, and counseling.
2. Comprehensive initial audiological assessments are imperative for high-quality hearing healthcare.
3. Neat and complete audiogram forms are necessary to reveal an audiologist's competent assessment and that all testing essential to that case was completed. Comparison of audiometric test results between clinicians and clinics is more accurate when the recorded results are thorough and clear.
4. It is the responsibility of the audiologist to evaluate any results that may be conflicting during behavioral testing and to use all audiometric and immittance measures available to confirm that test results are accurate.

Suggested Reading

American Speech-Language-Hearing Association (ASHA). Guidelines: Audiometric Symbols Committee on Audiologic Evaluation. www.asha.org/policy

American Speech-Language-Hearing Association (ASHA). Preferred practice patterns for the profession of audiology. www.asha.org/policy

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23 The Mystery of the 4000 Hz Air–Bone Gap

Kristi Oeding

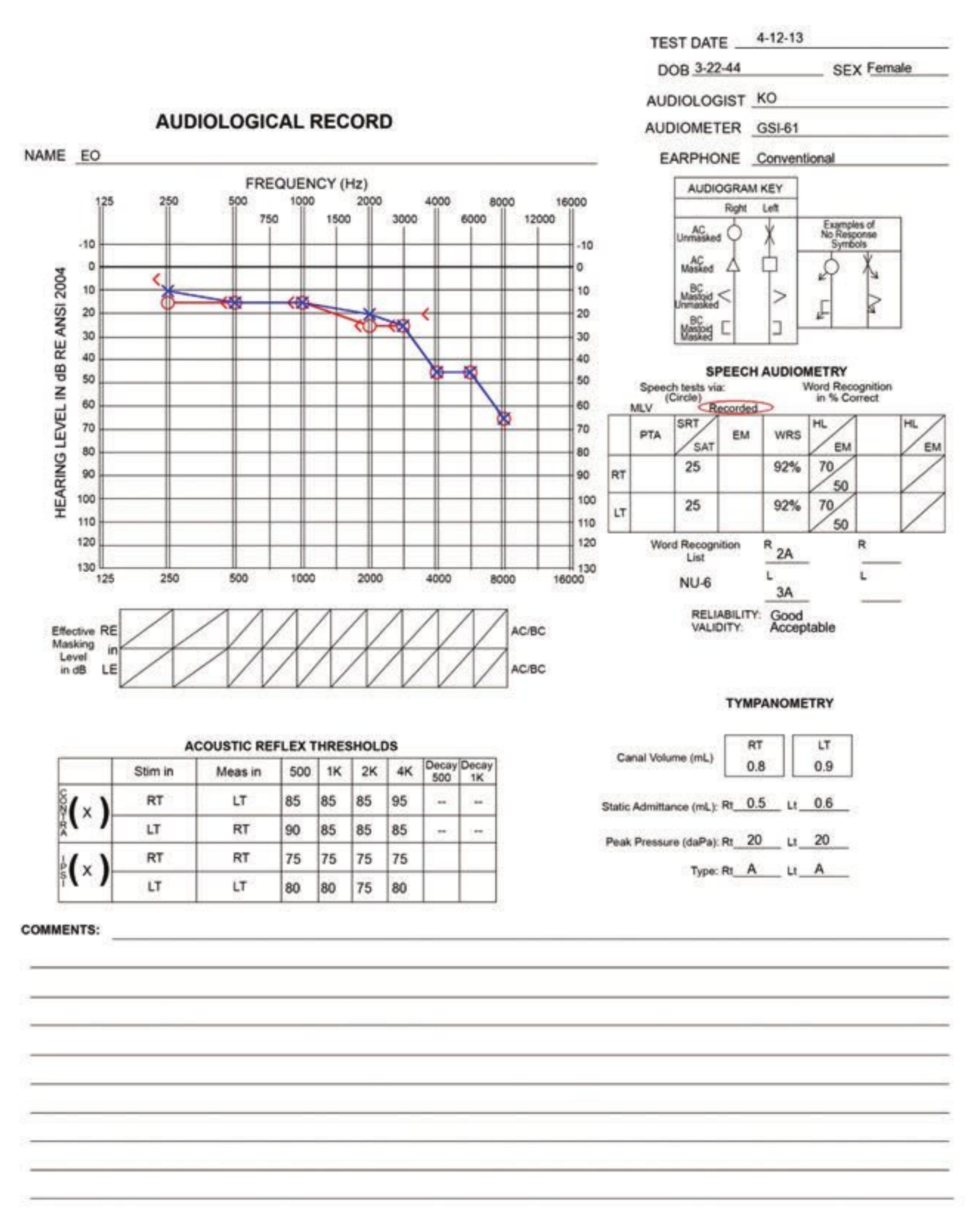
This case report describes the investigation of a patient with bilateral air–bone gaps at 4000 Hz and the possible causes for the presence of the air–bone gaps.

23.1 Clinical History and Description

EO is a 69-year-old female who arrived for an audiological examination prior to her appointment with an otologist. EO's primary complaint was dizziness that she described as a spinning and lightheaded sensation. EO reported she has had these symptoms for awhile, but the frequency of these symptoms has increased to every evening for the past month. The symptoms start at night and can sometimes wake her up in the morning. EO reported bilateral hearing loss, with the right ear being better than the left ear, and she notices difficulty communicating on the telephone using either ear. EO reported otalgia behind and in front of her left pinna, and the pain can radiate down her neck. EO reported that she occasionally has bilateral tinnitus and aural fullness, but this usually only occurs when she has sinus infections. No other otologic history or symptoms were reported.

23.2 Audiological Testing

Pure-tone air and bone conduction thresholds using TDH-50 headphones, speech recognition thresholds (SRTs), word recognition scores (WRSs), tympanometry, acoustic reflex thresholds (ARTs), and reflex decay were performed bilaterally (Fig. 23.1). The audiological examination revealed normal hearing from 250 to 1000 Hz, a slight sensorineural hearing loss from 2000 to 3000 Hz, and a moderate to moderately severe hearing loss from 4000 to 8000 Hz bilaterally. Prior to the introduction of masking noise at 4000 Hz due to the significant



TEST DATE 4-12-13

DOB 3-22-44 SEX Female

AUDIOLOGIST KO

AUDIOMETER GSI-61

EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME EO

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		25		92%	70 / 50		
LT		25		92%	70 / 50		

Word Recognition List NU-6 R 2A L 3A R ___ L ___

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	85	85	85	95	--	--
	LT	RT	90	85	85	85	--	--
IPSI (x)	RT	RT	75	75	75	75		
	LT	LT	80	80	75	80		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.8	0.9

Static Admittance (mL): Rt 0.5 Lt 0.6

Peak Pressure (daPa): Rt 20 Lt 20

Type: Rt A Lt A

COMMENTS:

Fig. 23.1 EO's comprehensive audiological and immittance examination using TDH-50 headphones.

(>10 dB HL) air–bone gap, a bilateral 25 dB air–bone gap was noted at 4000 Hz. SRTs revealed a slight loss in the ability to receive speech bilaterally. WRSs using the Northwestern University Auditory Test No. 6 (NU-6) word lists with a recorded female talker revealed normal ability (92%bilaterally) to recognize speech bilaterally. Immittance audiometry was performed and revealed normal tympanograms bilaterally, normal ARTs from 500 to 4000 Hz to ipsilateral and contralateral stimulation, and negative reflex decay at 500 to 1000 Hz bilaterally to contralateral stimulation.

23.3 Question to the Reader

1. What could cause the bilateral air–bone gap at 4000 Hz?

23.4 Discussion of Questions to the Reader

1. What could cause the bilateral air–bone gap at 4000 Hz?
   There are three possible causes for the bilateral air–bone gap at 4000 Hz:
   1. Mixed hearing loss—There is a possibility that EO has a conductive component to her hearing loss.
   2. False air–bone gap at 4000 Hz—There have been reports that false air–bone gaps are often present at 4000 Hz. In a recent report by Margolis et al (2013), the authors investigated the air–bone gap at 4000 Hz in patients with normal hearing and sensorineural hearing loss using a Radioear© B71 bone vibrator. Results from this study revealed an average air–bone gap of 10.6 dB HL in patients with normal hearing and an average air–bone gap of 14.1 dB HL in patients with sensorineural hearing loss. Of interest, the authors noted that as the magnitude of the air-conduction threshold increased (became poorer), the greater the measured air–bone gap. Pure-tone air conduction thresholds of 5 to 10 dB HL yielded an average air–bone gap of 10 dB HL, with a standard deviation (SD) of approximately 1 dB HL, whereas hearing thresholds greater than 60 dB HL revealed an average air–bone gap of 22 dB HL (SD approximately 7 dB HL). A hypothesis of why there was an increase in air–bone gaps based on hearing thresholds was due to possible changes in middle ear function from aging, but there was no correlation between age or air–bone gap at 4000 Hz.

      The authors suggested changing the reference equivalent threshold force levels (RETFLs = sound force level that corresponds to audiometric zero) to compensate/eliminate the false air–bone gap at 4000 Hz. The current RETFLs for 4000 Hz have a higher sensation level than REFTLs at 2000 Hz. Most of the other frequencies (250–2000 Hz) have a uniform − 12 dB per octave decrease in sensation level, but 3000 and particularly 4000 Hz increase instead of decrease in sensation level. The authors propose a − 12 dB per octave decrease at 3000 and 4000 Hz as well, which they hypothesize would eliminate the false air–bone gaps for sensorineural hearing losses, but as a result cause a potential negative air–bone gap for normal thresholds.
   3. Collapsed ear canals—The pressure of supra-aural earphones can cause the cartilaginous portion of the ear canal to collapse. When this occurs, a greater high-frequency hearing loss may occur than is really present. This can occur at all ages, but can be more prevalent in the older adult population. Some studies have reported a prevalence of 10 to 41%in persons over 60 years old. The average decline in hearing thresholds due to a collapsed canal was reported to range between 15 and 30 dB HL at one or more frequencies and typically occurs above 1000 to 2000 Hz. To compensate for the collapsed ear canal(s) an insert earphone can be used to keep the ear canal open.

23.5 Diagnosis and Recommended Treatment

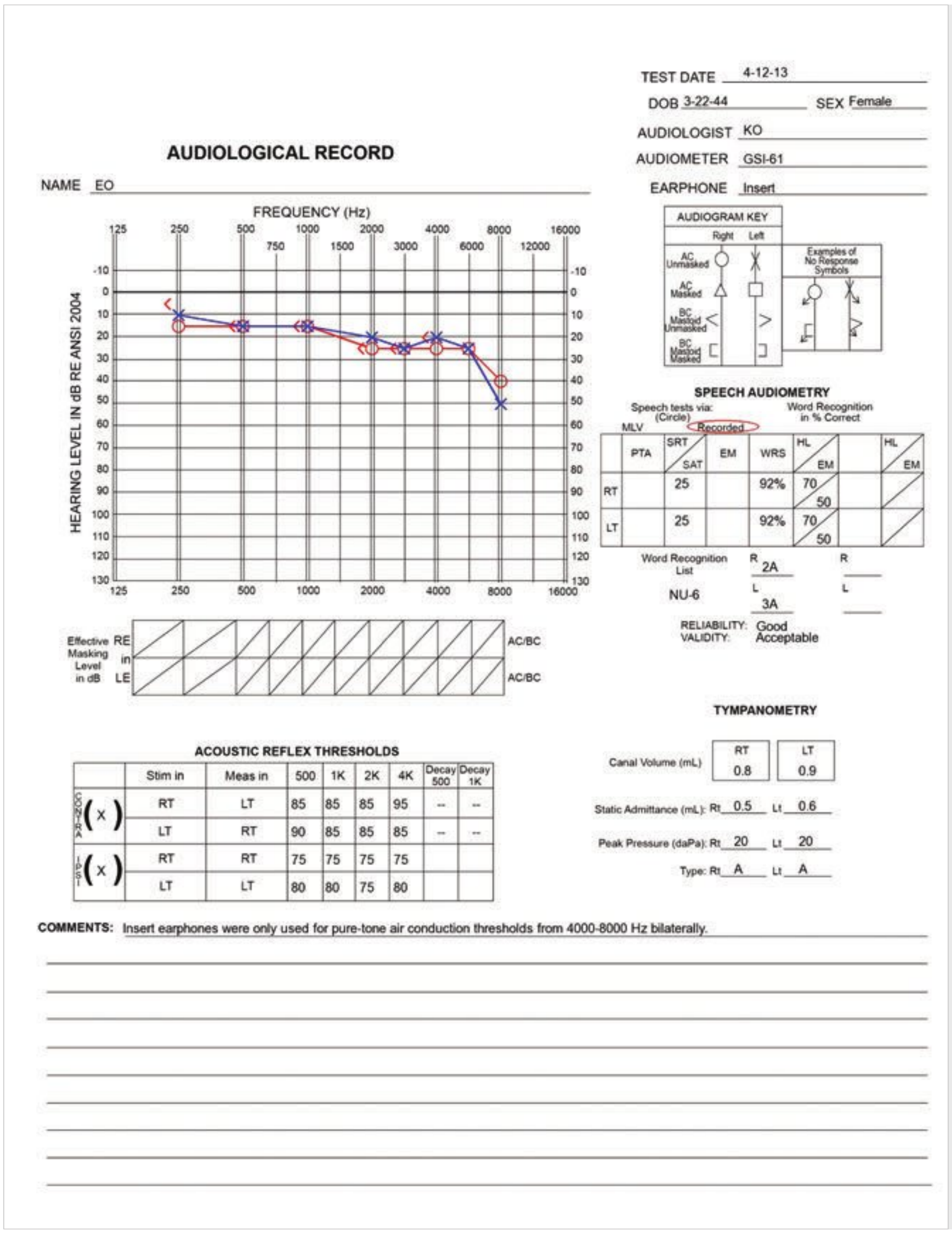
The author typically uses insert earphones prior to masking air–bone gaps at 4000 Hz to determine if a collapsed ear canal is the cause of the measured air–bone gap. Insert earphones (immittance tips) were used to retest air conduction thresholds from 4000 to 8000 Hz bilaterally (▯ Fig. 23.2).

23.6 Outcome

Results revealed an improvement in air conduction thresholds of 20 dB HL at 4000 and 6000 Hz and 25 dB HL at 8000 Hz in the right ear and 15 dB HL at 8000 Hz, 20 dB HL at 6000 Hz, and 25 dB HL at 4000 Hz in the left ear. As can be seen in this case report, the range of improvement using insert earphones ranged from 15 to 25 dB HL, which represents a significant improvement in the measured air conduction thresholds, and the results are in agreement with the immittance results. Another component to this is the standing wave at 6000 and 8000 Hz that occurs when headphones are used. One research study reported that standing waves from headphones could cause a decrease of 5 dB HL at 6000 Hz and 7 dB HL at 8000 Hz.

Some may argue that headphones should not be used for the audiological examination if inserts will prevent collapsing ear canals and standing waves. Insert earphones may overcome collapsed ear canals and standing waves, but if the seal is not tight, low frequencies may leak out of the ear canal, causing the appearance of a low-frequency conductive/mixed hearing loss. This can also be true for headphones if the headphones are not placed tightly around the pinna. There are advantages and disadvantages to using headphones, but insert earphones are important to have available for use to overcome the artifacts of collapsed ear canals, standing waves, and low-frequency leakage (i.e. if the headphones do not fit well on the patient’s head due to the anatomy).

Although there are many reasons for the presence of a “true” air–bone gap at 4000 Hz, in this case, the 4000 Hz air–bone gap was due to collapsing ear canals because the air–bone gap disappeared when the insert earphones were used. Also, if insert earphones had not been used, immittance examination results would have revealed normal middle ear function, indicating the air–bone gap may be due to collapsing ear canals. These results would warrant further exploration of the air–bone gap at 4000 Hz with insert earphones to rule out collapsing ear


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
Effective Masking Level in dB
RE
in
LE
AC/BC
AC/BC

AUDIOLOGICAL RECORD

TEST DATE 4-12-13
DOB 3-22-44 SEX Female
AUDIOLOGIST KO
AUDIOMETER GSI-61
EARPHONE Insert
NAME EO

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		25		92%	70 / 50		
LT		25		92%	70 / 50		

Word Recognition List NU-6 R 2A L 3A R ____ L ____

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL) RT 0.8 LT 0.9
Static Admittance (mL): Rt 0.5 Lt 0.6
Peak Pressure (daPa): Rt 20 Lt 20
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	85	85	85	95	--	--
	LT	RT	90	85	85	85	--	--
IPSI (X)	RT	RT	75	75	75	75		
	LT	LT	80	80	75	80		

COMMENTS: Insert earphones were only used for pure-tone air conduction thresholds from 4000-8000 Hz bilaterally.

Fig. 23.2 EO's comprehensive audiological and immittance examination using immittance tips for pure-tone air conduction thresholds from 4000 to 8000 Hz.

canals versus a false air–bone gap. If insert earphones (ER-3A, Etymotic Research, Inc., Elk Grove Village, IL, or immittance tips) are not available, a clinician could try repositioning the headphones so that the diaphragm aligns with the patient's ear canal with the headband tightened to pull the pinnae up, or place some earmold tubing in the ear canal to keep the ear canal patent.

23.7 Key Points

1. There are many reasons for an air–bone gap at 4000 Hz. It may be a "true" conductive component due to middle ear dysfunction, due to the current RETFLs for pure-tone bone conduction thresholds at 4000 Hz, or due to a collapsing ear canal.
2. An air–bone gap in the high frequencies, particularly at 4000 Hz, warrants further testing with insert earphones if headphones are used for pure-tone air conduction threshold testing to rule out the cause of the air–bone gap.

Suggested Reading

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Margolis RH, Eikelboom RH, Johnson C, Ginter SM, Swanepoel W, Moore BC. False air-bone gaps at 4 kHz in listeners with normal hearing and sensorineural hearing loss. Int J Audiol 2013; 52: 526–532

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24 Misuse of Insert Earphones during Pure-Tone Audiometry

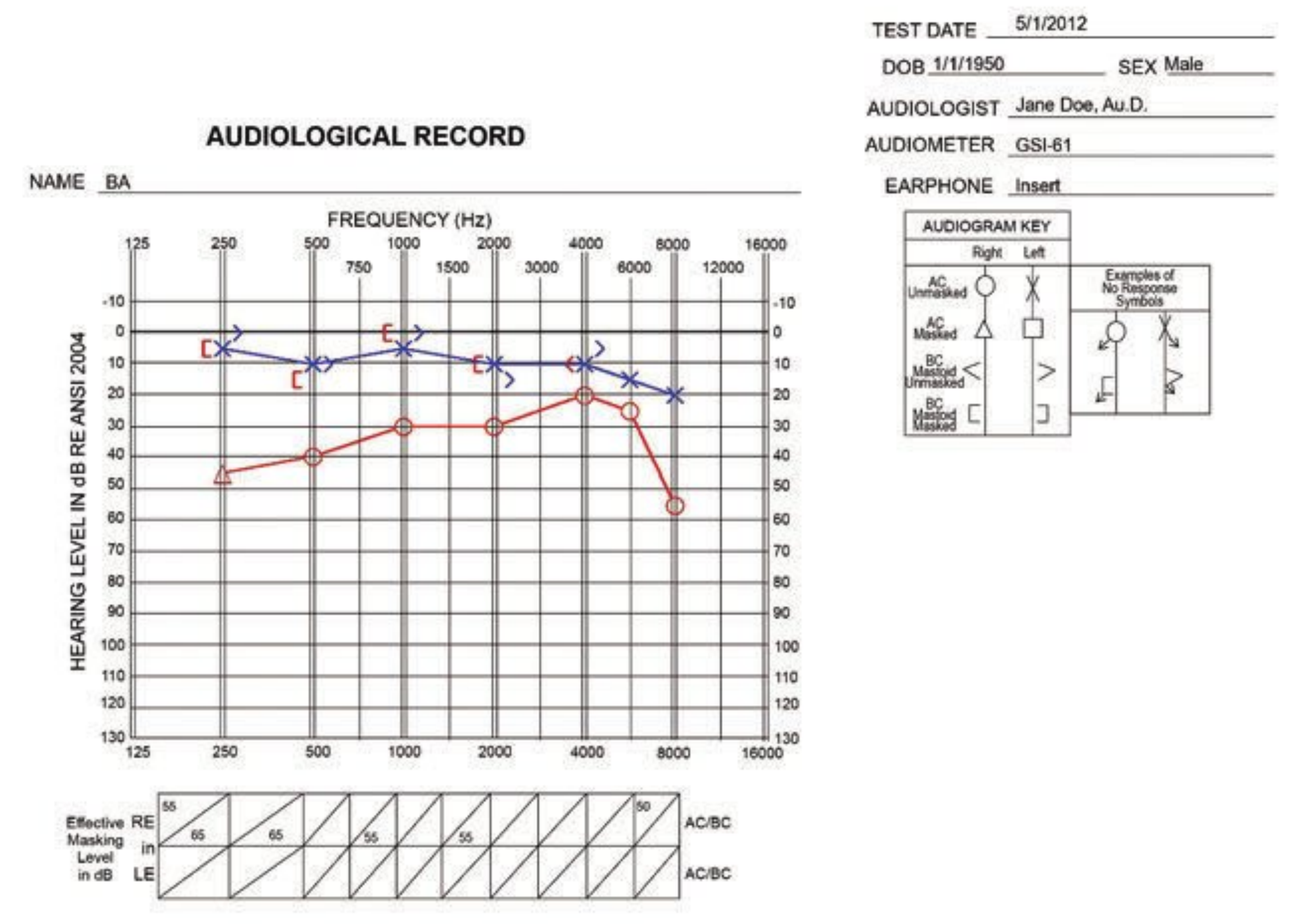
Daniel B. Putterman and M. Patrick Feeney

A 65-year-old male with serous otitis media (OM) in the right ear is seen for a series of audiological and otological examinations. The following case illustrates that when middle-ear pathology is present, such as a tympanic membrane (TM) perforation or a pressure-equalization (PE) tube placed in the TM, the use of insert earphones may exaggerate the extent of the low-frequency air conduction threshold. In addition to needlessly increasing patients' concern about their hearing status, this misinformation could affect how an otolaryngologist manages patient care.

24.1 Clinical History and Description

The patient is a 65-year-old male we will refer to as BA who has seen for a comprehensive audiological examination. The patient has a history of bilateral chronic OM that is greater in the right ear than the left ear. In recent years, BA has had normal hearing in the left ear and a mild to moderate conductive hearing loss (CHL) in the right ear. He has had repeated PE tubes inserted bilaterally, with the most recent surgery occurring several years ago, for which no audiological record is available. Most recently, BA has been followed by his otolaryngologist for OM in the right ear that has not resolved. BA reported that he has not had recent bouts with OM in the left ear. A PE tube was inserted in the right TM 7 months ago. The patient reported he had some minimal improvement in the hearing in his right ear following PE tube insertion.

Seven months ago, prior to the PE tube insertion, an audiologist at another facility completed an audiological examination on BA and then the audiologist completed an additional audiological examination 2 months after BA's otolaryngologist inserted the PE tube. Of note, the audiologist used ER-3A insert earphones (Etymotic Research, Inc., Elk Grove Village, IL) during audiological examination for both visits. The audiogram from the initial visit revealed a moderate CHL at 250 to 500 Hz, rising to a mild CHL at 1000 to 6000 Hz, and falling to a moderate hearing loss at 8000 Hz in the right ear (Fig. 24.1). There were significant air–bone gaps present at 250 to 3000 Hz. Immediately following this visit, the otolaryngologist inserted a PE tube into the TM of the right ear. When the patient was seen for follow-up 2 months post–PE tube insertion, the audiogram revealed even greater hearing loss in the right ear. Pure-tone thresholds at that visit revealed a moderately severe CHL at 250 Hz, rising to a moderate CHL at 500 Hz and rising further to a mild CHL at 1000 to 6000 Hz, then falling to a moderately severe hearing loss at 8000 Hz (Fig. 24.2). Significant air–bone gaps were once again present at 250 to 1000 Hz. Specifically, pure-tone thresholds at 250 and 500 Hz were 20 and 10 dB poorer, respectively, following the PE tube insertion. Tympanometric testing revealed an abnormally large ear-canal volume and no tympanometric peak, suggesting that the PE tube was functional, which was confirmed by BA's otolaryngologist. Therefore, despite the insertion of the PE tube to aerate the middle-ear space and prevent further buildup of fluid, the second audiogram revealed poorer low-frequency hearing when compared with the initial audiogram. The otolaryngologist described the preoperative and postoperative hearing results as "a paradoxical worsening of hearing." The recommended treatment was to follow BA to monitor his PE tube and hearing status in the right ear. BA arrived at our clinic for follow-up audiological and otological examination.



AUDIOLOGICAL RECORD

NAME BA

TEST DATE 5/1/2012

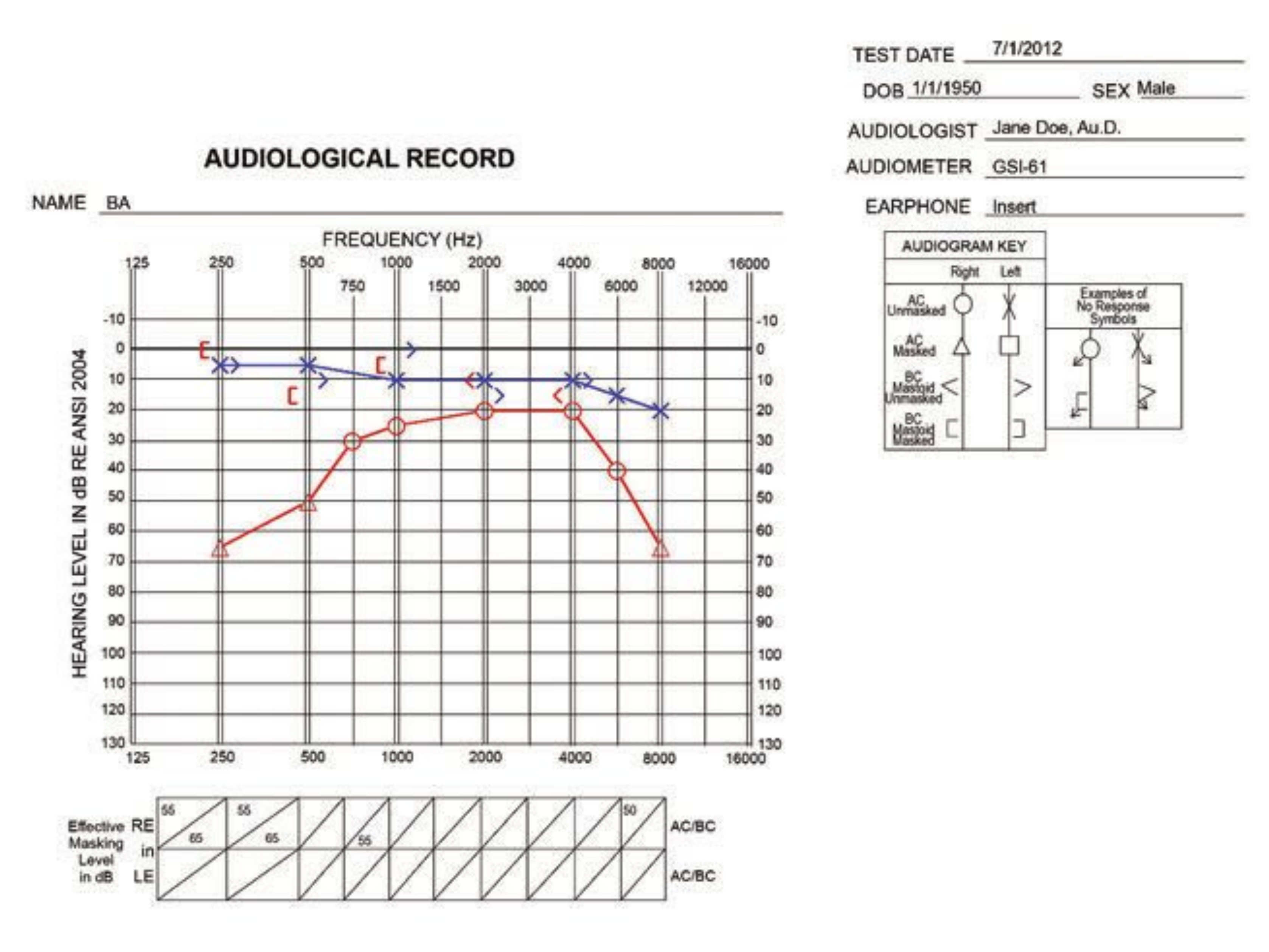
DOB 1/1/1950 SEX Male

AUDIOLOGIST Jane Doe, Au.D.

AUDIOMETER GSI-61

EARPHONE Insert

Fig. 24.1 Air and bone conduction thresholds (dB HL) before pressure-equalization tube placement in the right tympanic membrane. Air conduction thresholds were obtained using insert earphones.



AUDIOLOGICAL RECORD

NAME BA

TEST DATE 7/1/2012

DOB 1/1/1950 SEX Male

AUDIOLOGIST Jane Doe, Au.D.

AUDIOMETER GSI-61

EARPHONE Insert

Fig. 24.2 Air and bone conduction thresholds (dB HL) 2 months after pressure-equalization tube placement in the right tympanic membrane. Air conduction thresholds were obtained using insert earphones.

24.2 Audiological Testing

Prior to completing the audiological examination, the audiologist carefully inspected both ear canals via otoscopy. The audiologist noted the PE tube in the right TM and that it appeared to be patent. Immittance audiometry remains an integral portion of the audiological examination and, in conjunction with pure-tone and speech audiometry, aids an audiologist in evaluating the status of the middle ear and auditory pathway. Tympanometry and acoustic reflex thresholds (ARTs) were obtained (Fig. 24.3). Right ear tympanometry revealed a flat (Jerger type B) tympanogram with an abnormally large ear canal volume (2.4 mL). This would be an expected finding for an ear with an intact and patent PE tube. Ipsilateral and contralateral ARTs with the probe in the right ear were absent at 500, 1000, and 2000 Hz. Ipsilateral ARTs, with the probe and stimulus in the left ear, were present at 500, 1000, and 2000 Hz. These findings are consistent with PE tube placement in the right ear and normal hearing in the left ear with an intact tympanic membrane.

The audiologist completed pure-tone and speech audiometry using ER-3A insert earphones (Fig. 24.3). The pure-tone air conduction thresholds for the right ear were comparable to results from the audiogram completed with ER-3A insert earphones 2 months prior after PE tube placement. Left ear thresholds remained within normal limits with no significant change from the previous audiometric results. Speech recognition thresholds (SRTs) were obtained with recorded Central Institute of the Deaf (CID) W-2 spondaic words. SRTs were 25 dB HL in the right ear and 10 dB HL in the left ear. SRTs revealed a mild loss in the ability to receive speech in the right ear and normal ability to receive speech in the left. They were in good agreement with the three-frequency (500, 1000, and 2000 Hz) pure-tone averages for each ear, which were 30 and 8 dB HL for right and left ears, respectively. Word recognition scores (WRSs) were obtained with recorded 50-word CID W-22 phonetically balanced monosyllabic word lists at presentation levels of 70 dB HL to the right ear and 50 dB HL to the left ear and were found to be 96%bilaterally, as expected.

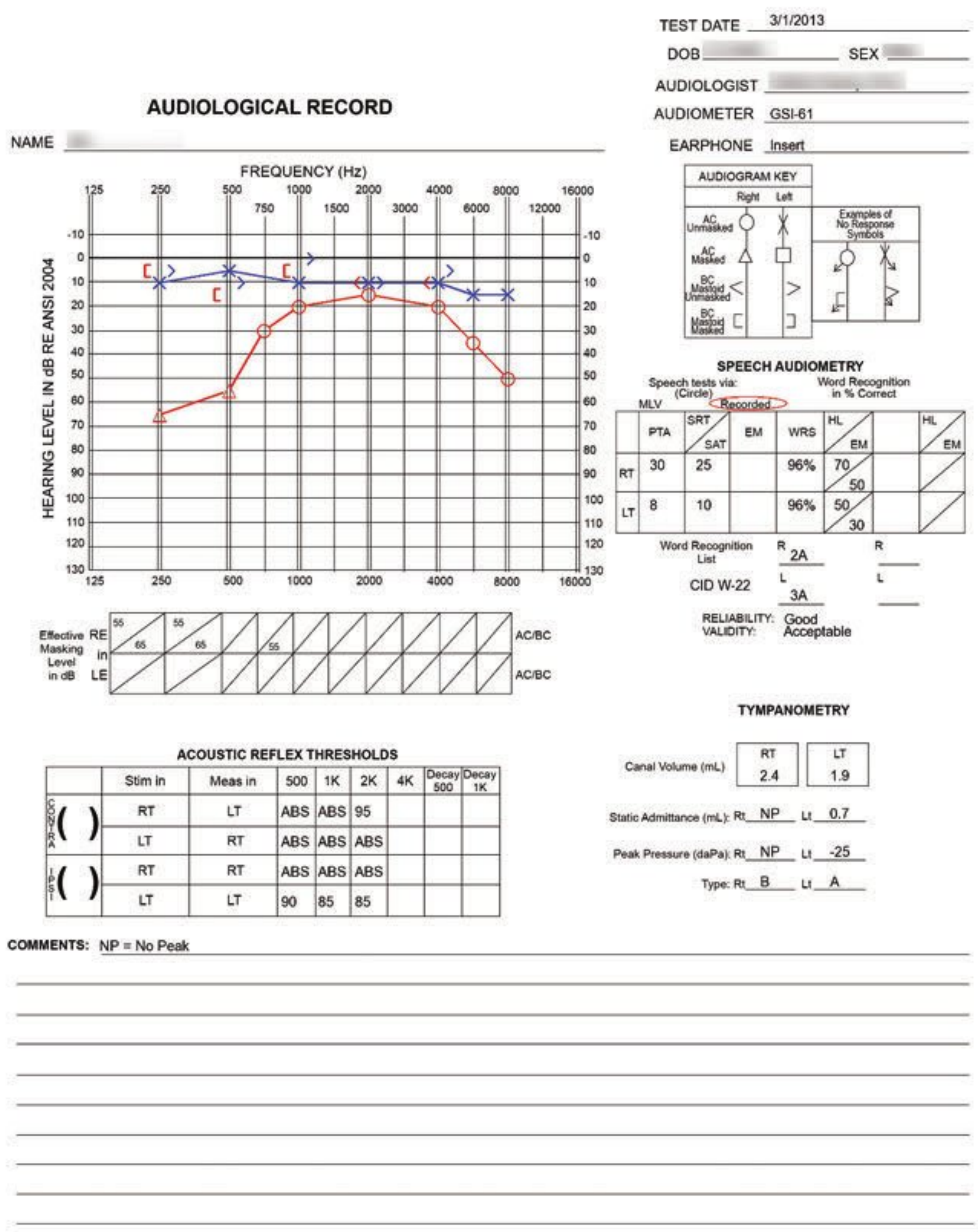
Next, the audiologist completed right ear pure-tone air conduction audiometry using TDH-50 P supra-aural headphones (Telephonics Corporation, Huntington, NY). Right ear low-frequency air conduction thresholds (Fig. 24.4) improved from those obtained with ER-3A insert earphones (Fig. 24.3). Specifically, there was a 15 dB improvement in thresholds at 250 and 500 Hz.

24.3 Questions to the Reader

1. What information, if any, must an audiologist consider prior to proceeding with an audiological examination?
2. What are some advantages and (equally important) disadvantages of using insert earphones during an audiological examination?
3. Why does this patient’s clinical history contraindicate the use of insert earphones?
4. What additional pathologies might contraindicate the use of insert earphones, and why?

24.4 Discussion of Questions to the Reader

1. What information, if any, must an audiologist consider prior to proceeding with an audiological examination?
   It is imperative that clinicians understand that an audiological examination should consistently involve critical thinking. Clinicians have many factors to consider prior to and during an audiological examination. The purpose of considering



TEST DATE 3/1/2013

DOB [redacted] SEX [redacted]

AUDIOLOGIST [redacted]

AUDIOMETER GSI-61

EARPHONE Insert

AUDIOLOGICAL RECORD

NAME [redacted]

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT	30	25		96%	70/50		
LT	8	10		96%	50/30		

Word Recognition List R 2A R

CID W-22 L 3A L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	2.4	1.9

Static Admittance (mL): Rt NP Lt 0.7

Peak Pressure (daPa): Rt NP Lt -25

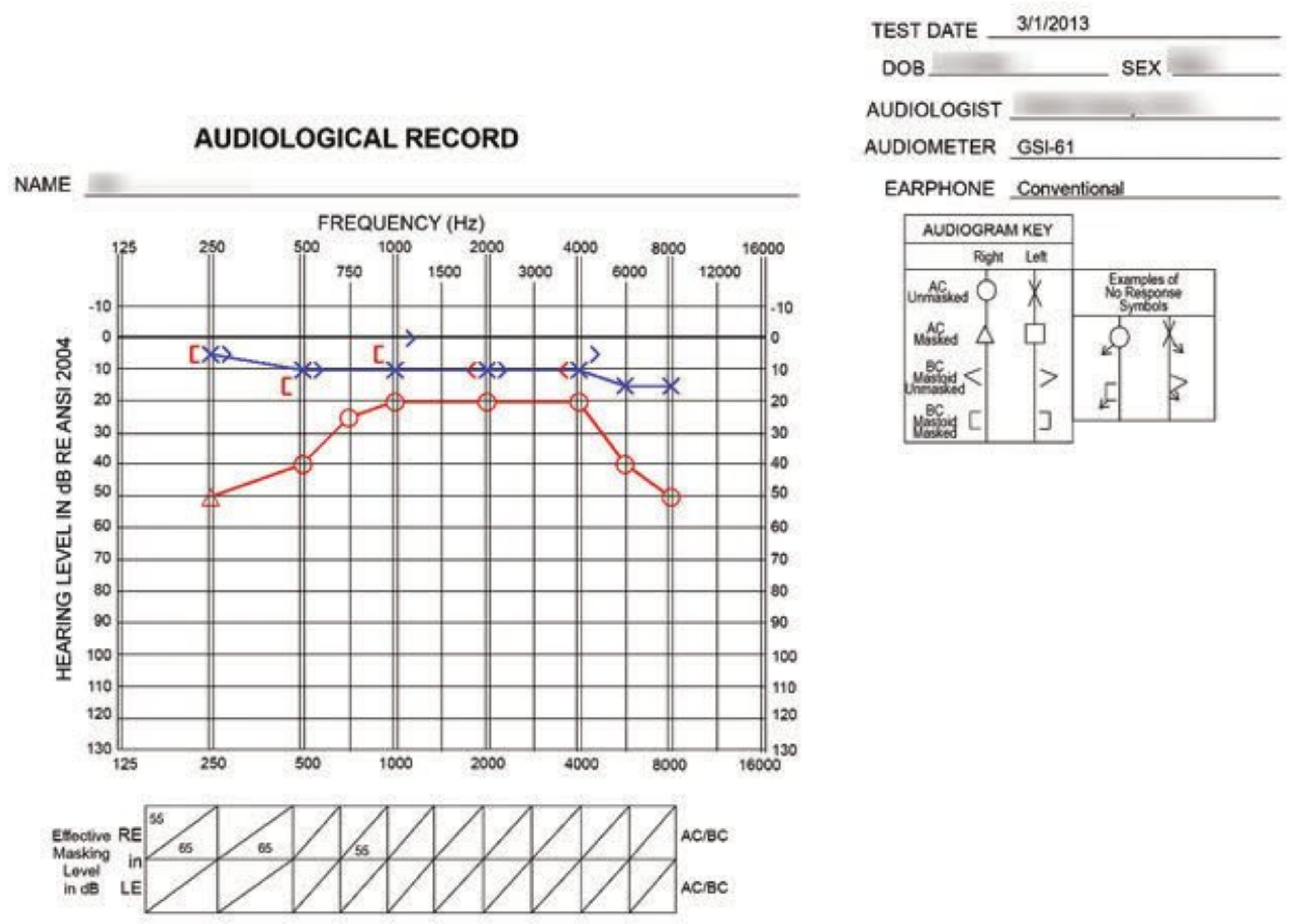
Type: Rt B Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	ABS	95			
	LT	RT	ABS	ABS	ABS			
IPSI ( )	RT	RT	ABS	ABS	ABS			
	LT	LT	90	85	85			

COMMENTS: NP = No Peak

Fig. 24.3 Air and bone conduction thresholds (dB HL) 7 months after pressure-equalization tube placement in the right tympanic membrane. Air conduction thresholds were obtained using insert earphones.



TEST DATE 3/1/2013

DOB [redacted] SEX [redacted]

AUDIOLOGIST [redacted]

AUDIOMETER GSI-61

EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME [redacted]

Fig. 24.4 Air and bone conduction thresholds (dB HL) 7 months after pressure-equalization tube placement in the tympanic membrane. Air conduction thresholds were obtained using supra-aural headphones.

these factors is to conduct the audiological examination in a manner that will (ideally) generate the most accurate results. These factors are inclusive of, but not limited to, age, language, language level, aural health history, anatomical abnormality, physical and/or sensory deficit, and mental health status. Medical record review, if available, as well as the interaction during patient intake and the case history can be most illuminating when identifying factors that may support adapting procedures during the audiological examination. In addition, otoscopy provides invaluable information concerning the status of the ear canal, TM, and occasionally the middle ear. Once these factors are assessed, the clinician must attempt to develop the best course of action with respect to test order, transducer type, and stimuli.

2. What are some advantages and (equally important) disadvantages of using insert earphones during an audiological examination?
   Insert earphones are useful for an audiological examination because they prevent the possibility of an acoustic leak, keep ear canals from collapsing, and provide greater interaural attenuation while lessening the occlusion effect as compared with supra-aural headphones. In addition, insert earphones may be advantageous for hearing aid fittings to obtain more accurate real-ear sound pressure level (SPL) measures due to a similar relative ear canal volume as when the patient is wearing a hearing aid. Conversely, if supra-aural headphones are not positioned appropriately over the entrance to the patient's ear canals, then there is likely to be acoustic leakage. Moreover, hearing thresholds at 6000 and 8000 Hz using supra-aural headphones can be inaccurate due to standing waves if the diaphragm of the headphone is situated adjacent to the ear canal entrance.

   It should, however, be noted that all of the advantages of insert earphones can be compromised by improper insert earphone placement. One way this can occur is if the foam tip is not fully inserted to the junction of the cartilaginous and osseous portions of the ear canal (full insertion depth). In addition, the foam tip must be compressed such that the foam does not expand in front of and block the opening to the transducer when it is inserted into the ear canal. Gently pulling back on the pinna while asking the patient to open his or her jaw will straighten the ear canal, which will make it easier to fully insert the foam tip. Another potential problem is that the insert earphone opening can also be easily plugged by debris such as cerumen and drainage that may be present in the ear canal. Therefore, use of an insert earphone would not be prudent when examining patients with cerumen partially occluding the ear canal or patients with draining ears.

   Hygiene and patient comfort may or may not be an advantage for either transducer. Insert earphones may appear to be more hygienic because the foam tips should be discarded after a single use. This is only effective if the clinician is conscientious about removing and discarding the tips after seeing each patient while wearing gloves or practicing meticulous hand hygiene. Supra-aural headphones should be disinfected with wipes after each use, but they can also be fit with one-time-use covers that are designed to be acoustically transparent. In reality, the expense to the clinic of disposable gloves, foam tips, and headphone covers may determine whether some or all of these options are available to the clinician. Comfort is relative to the patient; some patients may dislike the feeling of having something inserted into their ear canals, whereas other patients may dislike the tight fit of headphones positioned on their head.
3. Why does this patient's clinical history contraindicate the use of insert earphones?
   With regard to pure-tone audiological practice, there are probably some inaccurate assumptions made by even the most experienced clinicians. Specifically, some clinicians believe that as long as equipment is properly calibrated, the supra-aural and insert transducers will yield equivalent SPLs. As a result, clinicians may not consider that middle-ear impedance, which can be altered by middle-ear pathologies, can impact the SPL delivered to the cochlea by either transducer.

   According to Voss et al, the SPL generated by insert earphones (or supra-aural headphones) will vary even in normal adult ear canals. Specifically, there can be a standard deviation as great as 4 dB at test frequencies above 200 Hz, inclusive of all of the conventionally tested audiometric frequencies (250 to 8000 Hz). This variability is small, however, compared with the additional findings from Voss et al when the external or middle ear is abnormal, especially with low-impedance pathologies. For example, when a patient like BA has a PE tube, the low-frequency (200–400 Hz) SPL that is generated in the ear canal can be 5 to 25 dB less than that of a normal ear when insert earphones are used. In contrast, supra-aural headphones are much less affected than insert earphones. Therefore, using insert earphones to examine patients with PE tubes can result in air conduction thresholds that are significantly exaggerated in the low frequencies, yielding greater than "true" air–bone gaps.
4. What additional pathologies might contraindicate the use of insert earphones, and why?
   Voss et al also reported how SPL fluctuates with other ear pathologies relative to normal ears when one is using insert earphones and supra-aural headphones. As might be expected, when a patient has a pinhole perforation of the TM, the results are similar to when a PE tube is placed in the TM. This suggests that insert earphones are again more likely to provide misleading low-frequency air conduction results than would supra-aural headphones.

   When there is a mastoid bowl, however, the results are less predictable as to which transducer should be used. A mastoid bowl is a depression resulting from the absence of mastoid air cells after they have been drilled out by an otolaryngologist. This procedure is known as a mastoidectomy, and typically these air cells are removed because they have become infected. Mastoid bowls generate an SPL that is 5 to 30 dB lower than measured in normal ears above 2000 Hz, and consistently 5 to 15 dB lower than normal ears below 2000 Hz when insert earphones are used. Voss et al reported that the SPL when using supra-aural headphones was 5 to 25 dB lower than normal ears below 500 Hz, and that their use generated sharp increases and decreases in SPL relative to normal ears between 1000 and 4000 Hz. Additionally, larger TM perforations yielded results that were similar to the results obtained for mastoid bowls.

24.5 Diagnosis and Recommended Treatment

As was stated in the clinical history and description section of this case report, BA had a history of recurring OM. His otolaryngologist had attempted to resolve a recurrence of serous OM in the right ear over the past 7 months. Based upon his progress note, the otolaryngologist was perplexed when the results from the audiological examination completed 2 months after placement of the PE tube in the right TM revealed significantly greater air–bone gaps at 250 to 500 Hz than the audiological examination completed prior to inserting the PE tube.

Given that hearing thresholds at 250 to 500 Hz in the right ear had become poorer, BA's otolaryngologist may have become concerned that the OM had caused additional middle-ear complications, such as impacting the structure of the middle-ear ossicles. Moreover, BA's otolaryngologist may have suspected the onset of a cholesteatoma or otosclerosis. If the otolaryngologist had elected to become more aggressive in his treatment plan for BA, he may have ordered imaging or considered an exploratory procedure. Fortunately, in this case the otolaryngologist adopted a conservative "wait-and-see" approach. The potential for serious mismanagement of this patient should, however, highlight how important it is for clinicians to realize that transducers may not provide equal results across patients when the air space of the ear canal and middle ear are abnormal and/or the surface of the TM is perforated.

Because the second audiologist considered this possibility, the otolaryngologist was provided more accurate information about the hearing sensitivity of the right ear. Therefore, additional middle-ear complications were no longer considered by BA's otolaryngologist, and he continued to treat BA's OM with antibiotics while advising BA to keep his ear canal dry. The otolaryngologist noted, after stereomicroscopy, that there no longer appeared to be drainage present in BA's right ear canal.

24.6 Outcome

The outcome for BA is that, through otological intervention, BA's right ear serous OM appears to have resolved. Of equal importance, BA was not unnecessarily treated as if additional middle-ear complications were present. This case report provides an excellent example of what may be a common cause of inaccurate audiometric results when assessing low-frequency thresholds using insert earphones.

Audiologists frequently examine patients who, for one reason or another, have an abnormal ear canal and/or middle-ear volume. Frequently, this is due to the presence of TM perforation, PE tube placement, or mastoidectomy. Small TM perforations can often elude clinicians during otoscopy, which is another reason why immittance audiometry plays an important role in diagnosis. When insert earphones are used to test patients with these pathologies then hearing thresholds could be overestimated, as was initially the case for BA.

24.7 Key Points

1. Although there are certainly advantages to using insert earphones for audiological examination, there are disadvantages to their use as well.
2. Calibrated supra-aural headphones and insert earphones alike are not immune to variability in generating different intensity levels across patients because interpatient differences in ear-canal and middle-ear volume can alter the SPL before the sound energy reaches the cochlea.
3. Audiologists should strongly consider using supra-aural headphones in lieu of insert earphones during audiological examination of patients with low-impedance middle-ear problems, and particularly those with PE tubes or small perforations of the TM.

24.8 Acknowledgments

The authors wish to acknowledge support from VA RR&D Center Award C9230C. The contents of this case do not represent the views of the Department of Veterans Affairs or the United States Government.

Suggested Reading

Voss SE, Rosowski JJ, Merchant SN, Thornton AR, Shera CA, Peake WT. Middle ear pathology can affect the ear-canal sound pressure generated by audiologic earphones. Ear Hear 2000; 21: 265–274

Voss SE, Herrmann BS. How does the sound pressure generated by circumaural, supra-aural, and insert earphones differ for adult and infant ears? Ear Hear 2005; 26: 636–650

Wilber LA. Calibration: transducers for audiologic testing. In: Katz J, ed. Handbook of Clinical Audiology. Baltimore, MD: Lippincott Williams and Wilkins; 2002:88–95

25 Pseudohypoacusis: The Stenger Test

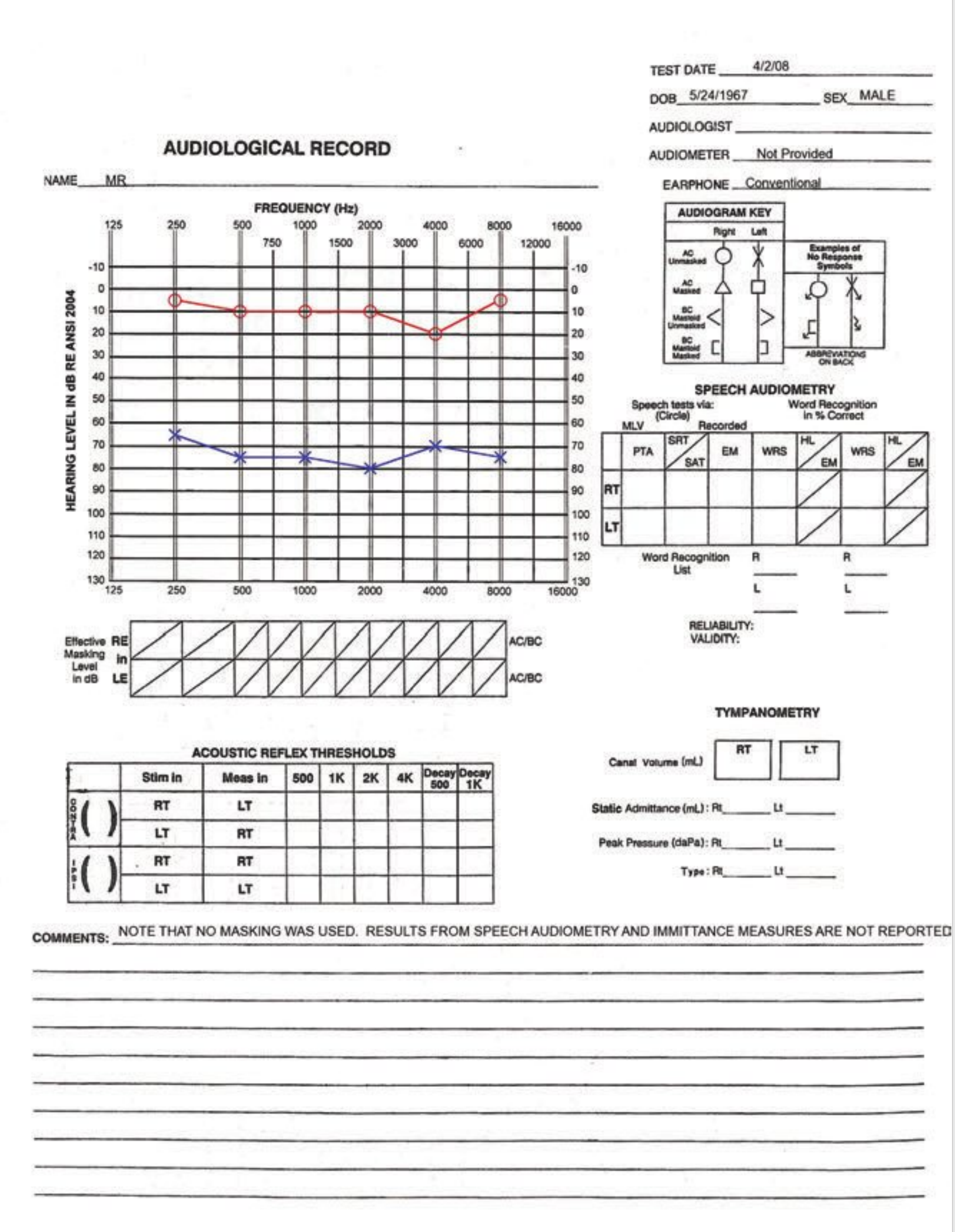
Ross J. Roeser

Some authors use the term pseudohypacusis, rather than pseudohypoacusis. Either spelling is correct, because hyp means, “below, beneath, or under,” such as in hypochondriac; or “less than normal or deficient,” and such as in hypofunction (note that in both examples hyp is followed by an o). The author of this case report uses pseudohypoacusis because it is felt to be more accurate.

A 46-year-old male with a history of significant physical trauma to the left side of his head presents with pure-tone findings from two previous audiological examinations completed within 4 months of each other at another clinic revealing hearing within normal limits in the right ear with the exception of a slight hearing loss at 4000 Hz and a fluctuating severe to profound hearing loss in his left ear. The audiological examination performed by the author of this case report, however, did not agree with the previous findings at the other clinic. The results of the audiological examination completed by this author do not support the severity of the hearing loss in the left ear that was reported on the two previous evaluations, suggesting pseudohypoacusis.

25.1 Clinical History and Description

The patient was a 46-year-old male referred by his attorney. The patient’s primary complaint was hearing loss in his left ear following an occupational injury. Approximately 1 year prior to the audiological examination the patient stated he was relocating a barrel at his employment setting when he reported that the barrel “exploded.” The left side of his head was near the barrel and the resulting blast caused trauma to the left side of his face that required two plastic surgeries. Prior to the audiological examination, the patient was seen on two different occasions in a general practitioner’s office for pure-tone audiometry. The results are reported in Fig. 25.1 and Fig. 25.2. It is clear



AUDIOLOGICAL RECORD

TEST DATE 4/2/08
DOB 5/24/1967 SEX MALE
AUDIOLOGIST
AUDIOMETER Not Provided
EARPHONE Conventional
NAME MR

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT							
LT							

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

TYMPANOMETRY

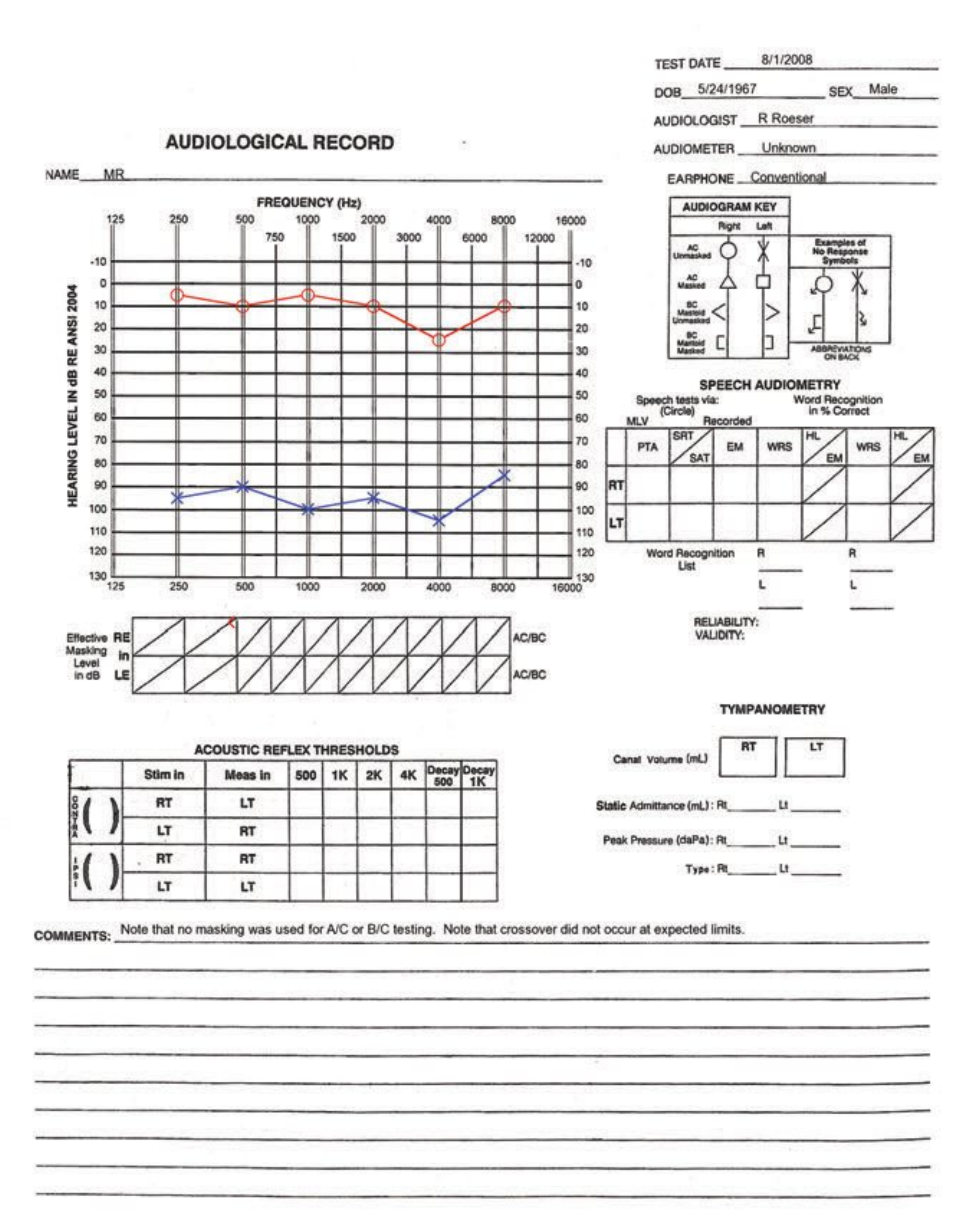
Canal Volume (mL) RT LT
Static Admittance (mL): Rt_______ Lt_______
Peak Pressure (daPa): Rt_______ Lt_______
Type: Rt_______ Lt_______

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: NOTE THAT NO MASKING WAS USED. RESULTS FROM SPEECH AUDIOMETRY AND IMMITTANCE MEASURES ARE NOT REPORTED

Fig. 25.1 Initial pure-tone audiometry results obtained at the patient’s general practitioner’s office prior to the diagnostic audiological evaluation. Note that no masking was used, and results from speech and immittance audiometry were not reported.



AUDIOLOGICAL RECORD

NAME MR

TEST DATE 8/1/2008

DOB 5/24/1967 SEX Male

AUDIOLOGIST R Roeser

AUDIOMETER Unknown

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT							
LT							

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt____ Lt____

Peak Pressure (daPa): Rt____ Lt____

Type: Rt____ Lt____

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: Note that no masking was used for A/C or B/C testing. Note that crossover did not occur at expected limits.

Fig. 25.2 Second pure-tone audiometry results obtained at the patient's general practitioner's office. The same audiometric limitations noted for the initial audiological examination are seen for these results (Fig. 25.1).

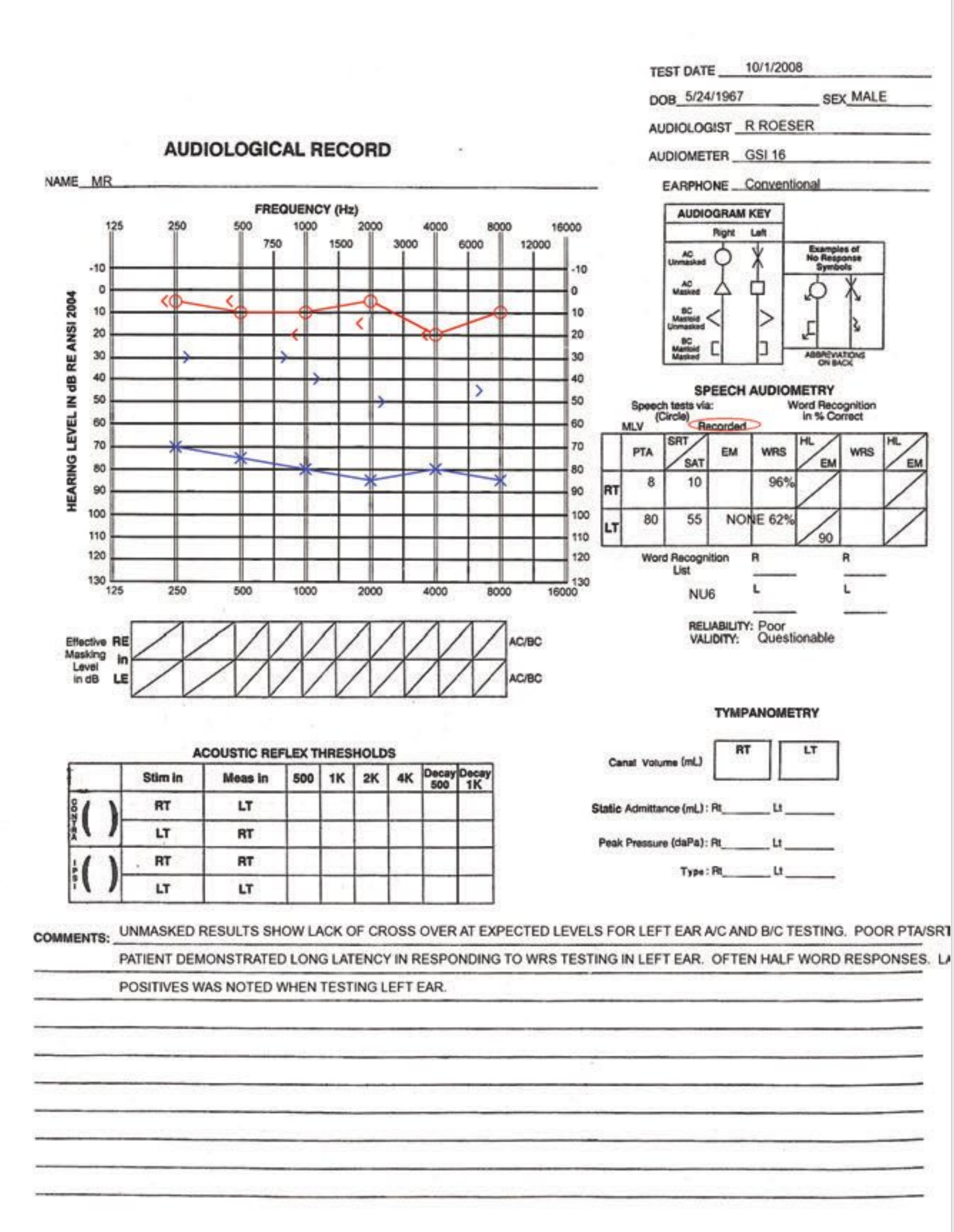
that the individual performing the audiological tests shown in Fig. 25.1 and Fig. 25.2 was unfamiliar with diagnostic audiological testing due to the lack of needed masking for air conduction (AC) thresholds. In addition, speech and immittance audiometry was not reported. Comparison of pure-tone thresholds in Fig. 25.1 and Fig. 25.2 shows poor test–retest reliability, which in turn questions the validity of these findings.

During the case history, the patient denied any hearing loss prior to the accident. Following the accident, the patient stated he had constant high-frequency tinnitus and was experiencing significant difficulty communicating when in a noisy listening environment.

25.2 Audiological Testing

Initial results from pure-tone and speech audiometry by an audiologist in the author's clinic is reported in Fig. 25.3. Several discrepancies in the patient's responses during this initial examination were noted. First, unmasked left AC thresholds (75–85 dB HL) using supra-aural earphones demonstrated no crossover at the expected hearing levels. Crossover for AC presented signals using supra-aural earphones should occur at 60 to 65 dB HL above the BC threshold of the nontest ear. If this were the case, then initial responses in this range should have occurred for this patient. Second, unmasked left ear BC thresholds revealed no crossover at the expected hearing level. Crossover for BC-presented signals to the left ear should essentially match the BC threshold of the better-hearing right ear, but instead are 20 to 35 dB poorer than the BC thresholds of the better right ear. Third, the speech recognition threshold (SRT) for the left ear (55 dB HL) was significantly better than what would be predicted from the pure-tone thresholds. The SRT should be within 5 dB of the pure-tone average (PTA) at 500, 1000, and 2000 Hz, but instead is 25 dB better than the PTA.

As shown in Table 25.1, the pure-tone Stenger test was performed at 500 Hz and 4000 Hz. Note that the patient responded (+) at 500 and 4000 Hz when the stimuli were presented only to his right ear at 10 dB above his voluntary thresholds at 500 and 4000 Hz. He failed, however, to respond when the stimuli were presented only to his left ear at 10 dB below his voluntary threshold or when the stimuli were presented bilaterally at 10 dB below his voluntary threshold in his left ear and simultaneously at 10 dB above his voluntary threshold of his right ear.



AUDIOLOGICAL RECORD

NAME MR

TEST DATE 10/1/2008
DOB 5/24/1967 SEX MALE
AUDIOLOGIST R ROESER
AUDIOMETER GSI 16
EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	8	10		96%			
LT	80	55	NONE	62%	90		

Word Recognition List NU6 R ____ L ____ R ____ L ____

RELIABILITY: Poor
VALIDITY: Questionable

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt____ Lt____
Peak Pressure (daPa): Rt____ Lt____
Type: Rt____ Lt____

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: UNMASKED RESULTS SHOW LACK OF CROSS OVER AT EXPECTED LEVELS FOR LEFT EAR A/C AND B/C TESTING. POOR PTA/SRT
PATIENT DEMONSTRATED LONG LATENCY IN RESPONDING TO WRS TESTING IN LEFT EAR. OFTEN HALF WORD RESPONSES. LA
POSITIVES WAS NOTED WHEN TESTING LEFT EAR.

Fig. 25.3 Unmasked audiometric results obtained for the patient by the author of this case report.

In this case, there is no reason for the patient not to respond to the stimuli in his right ear when bilateral presentation was complete. The Stenger phenomenon, however, resulted in the patient reporting he was unable to perceive the stimulus only in his right ear when both ears were stimulated. Note that when a patient does respond to the initial bilateral stimulation during the Stenger test, the response is recorded as a negative Stenger (i.e., does not support the presence of pseudohypoacusis), but when the patient does not respond to the bilateral stimulation (+10 dB sensation level [SL] in the better ear and –10 dB SL in the poorer ear) the response is recorded as a positive Stenger (i.e., supports the presence of pseudohypoacusis).

Upon completion of the Stenger test, the minimum contralateral interference levels (MCILs) were measured for 500 and 4000 Hz. Results are shown in Table 25.2. Note that the patient failed to respond when the stimuli were presented above threshold in the right ear at 10 dB SL until the pure-tones in his left ear were decreased to at 30 dB HL in his right ear and 45 dB HL in his left ear. The MCIL procedure allows the audiologist to predict that "true" threshold is 10 to 15 dB below the stimulus level at which the patient responded voluntarily. In this case, the MCIL would predict that the "true" threshold at the test frequency is 15 to 20 dB HL for 500 HZ and 30 to 35 dB HL for 4000 Hz. This finding would suggest that the hearing level in the left ear at the two test frequencies is no greater than a mild hearing loss. At this point, the patient was referred for additional audiological tests (see below) confirming the results of the Stenger test and MCIL.

Table 25.1 Results from the pure-tone Stenger test

500 Hz			4000 Hz		
Pure-tone in the right ear (dB HL)	Pure-tone in the left ear (dB HL)	Patient response	Pure-tone in the right ear (dB HL)	Pure-tone in the left ear (dB HL)	Patient response
20	–	+	30	–	+
–	65	–	–	75	–
20	65	–	30	75	–

Table 25.2 Results from the minimum contralateral interference level test

500 Hz			4000 Hz		
Pure-tone in the right ear (dB HL)	Pure-tone in the left ear (dB HL)	Patient response	Pure-tone in the right ear (dB HL)	Pure-tone in the left ear (dB HL)	Patient response
20	–	+	30	–	+
20	65	−	–	75	−
20	60	−	30	75	−
20	60	−	30	70	−
20	55	−	30	65	−
20	50	−	30	60	−
20	45	−	30	55	−
20	40	−	30	50	−
20	35	−	30	45	+
20	30	+			

25.3 Questions to the Reader

1. What is the Stenger phenomenon?
2. Pseudohypoacusis literally means false (pseudo), reduced hearing (hypoacusis). Is this the best term to use when referring to patients who are found to be uncooperative on behavioral audiometric tests?
3. What process should an audiologist follow when a patient reveals discrepancies in the results of his or her audiological examination?
4. How should an audiologist communicate with a patient who is thought to be pseudohypoacusic?
5. What specific procedures are available for uncooperative patients during conventional audiological examinations?
6. What additional procedure(s) should be considered for patients thought to be pseudohypoacusic?

25.4 Discussion of Questions to the Reader

1. What is the Stenger phenomenon?
   The Stenger phenomenon occurs when ears are stimulated bilaterally and one ear is presented with an SL that is higher than the SL to the opposite ear. In such cases, only the ear with the higher SL (the louder) will perceive the stimulus. For example, if hearing is normal bilaterally and one ear is presented with a stimulus at 55 dB HL and the other at 20 dB HL, the patient will only perceive the stimulus in the ear with the stimulus at 55 dB HL.
2. Pseudohypoacusis literally means false (pseudo), reduced hearing (hypoacusis). Is this the best term to use when referring to patients who are uncooperative on behavioral audiometric tests?
   Historically, the term malingering was used with patients who were thought to be feigning results on audiological examinations. This term, however, is used in the legal arena to imply that individuals are purposefully not cooperating because they are seeking to gain, mostly for monetary payment for an injury that is not present. Because the specific intent for which the patient does not comply with testing cannot be accurately determined the term malingering should be avoided. Other terms, such as nonorganic hearing loss and functional hearing loss have replaced malingering by implying that there is no scientific or logical basis for the results of the audiological examination. Pseudohypoacusis correctly describes this process and is now the preferred term.
3. What process should an audiologist follow when a patient reveals discrepancies in the results of his or her audiological examination?
   An audiologist should try to avoid an embarrassing mistake by concluding the patient is at fault when test results do not agree when in fact there could be legitimate reasons that audiological results do not agree. If this occurs, audiologists should follow three basic steps:
   a) First, the audiologist should be sure proper procedures were followed and question if the patient was instructed properly? Did the patient know how to respond?
   b) Second, the audiologist should be sure the equipment is functioning properly. Is the equipment turned on? Has the equipment been recently calibrated? Are the input/output switches in the proper positions?
   c) Finally, the audiologist needs to check the patient. Did the patient understand the instructions?
   If these steps are correct and the patient continues to provide discrepant test results, then it is justified to conclude that the patient is displaying pseudohypoacusic behavior.
4. How should an audiologist communicate with a patient who is thought to be pseudohypoacusic?
   When pseudohypoacusis is suspected and confirmed, there are two approaches to follow. One approach is the “easy way out” and the other approach is “authoritarian.” Each audiologist will need to decide which approach is felt to be best for each patient and for himself/herself. The easy way out approach involves informing the patient that “something must be wrong, because test results are not in agreement,” and the results suggest that hearing may be better than what the patient is volunteering. The patient is then asked if he or she knows what might be wrong and that it may be that he or she has misunderstood the instructions concerning when to respond. It is reemphasized that the patient must respond, “even when sounds are just audible.” The audiologist might add that there are other tests that can be performed that require the patient to be “put to sleep,” and the use of these additional tests can be avoided if the results are more consistent. The patient is then provided with another

Table 25.3 Tests specifically developed for pseudohypoacusis

Procedure	Type of test
Test–retest threshold reliability	Qualitative
SRT/PTA agreement	Qualitative
Failure to demonstrate a shadow curve	Qualitative
Stenger test for pure-tone and speech	Quantitative
Doerfler-Stewart test	Qualitative
Lombard reflex	Qualitative
Delayed auditory feedback	Qualitative
Swinging story	Qualitative
Immittance measures—ARTs	Quantitative

Abbreviations: ART, acoustic reflex threshold; PTA, pure-tone average; SRT, speech recognition threshold.
Source: Adapted from Shoup and Roeser, 2007.
Note: ARTs are indicative of pseudohypoacusis when pure-tone ARTs are obtained at hearing levels below voluntary thresholds. It is not possible to predict pure-tone thresholds from ARTs elicited from pure-tones. Use of broad or narrow band noise has been used to predict pure-tone thresholds from ARTs. Evoked response audiometry (ABRs and middle latency responses [MLRs]) and otoacoustic emissions (OAEs) could also be considered but are used for diagnostic audiological procedures for patients other than pseudohypoacusic patients.

opportunity to repeat the test, and hopefully the patient will become more cooperative.

The authoritarian approach is sterner. The patient is counseled that based on the initial test results there is evidence that he or she is not cooperating and that the patient is "wasting the audiologist's time." The patient is then counseled that he or she can either be more cooperative or the test session will be terminated and a report prepared and forwarded to the referring source indicating the test results are not valid. As one last chance, the patient is asked to try harder and the tests will be repeated if he or she agrees to provide more valid and consistent responses.

5. What specific procedures are available for uncooperative patients during conventional audiological examinations?
   Table 25.3 lists a number of behavioral procedures developed specifically for the audiological examination of pseudohypoacusic patients. Note that most of the pseudohypoacusic test procedures are qualitative, rather than quantitative. The exception is the Stenger test. Unfortunately the Stenger test can only be used when patients demonstrate asymmetric or unilateral hearing loss with threshold differences of at least 35 to 40 dB between ears.

   One critically important modification to the conventional clinical protocol for obtaining pure-tone and speech thresholds is that the conventional Hughson-Westlake (descending/ascending) method is not used. Only an ascending approach should be used for patients thought to be pseudohypoacusic. The rationale for this modification is that if using the Hughson-Westlake procedure, the patient would be provided a "loudness yardstick" that will be used as a psychological reference if suprathreshold levels are initially presented. Using only an ascending threshold method by carefully and slowly increasing the intensity level and waiting for the patient to respond helps prevent establishing a "loudness yardstick" at high suprathreshold presentations.
6. What additional procedures should be considered for patients thought to be pseudohypoacusic?
   More advanced audiological procedures include acoustic reflex thresholds (ARTs) as part of immittance audiometry, distortion-product otoacoustic emissions (DPOAEs), and auditory brainstem response (ABR) or middle latency response (MLR) thresholds.

25.5 Additional Testing

Despite repeated attempts to define hearing thresholds in the left ear that were more valid, the patient continued to fail to cooperate. An immittance test battery, DPOAEs, and an ABR test were performed. Each of these procedures failed to document the severity of the voluntary left ear behavioral thresholds. The results of these additional tests supported the findings from the MCIL segment of the Stenger test.

25.6 Diagnosis and Recommended Treatment

It was found that the patient had hearing within normal limits in the right ear and a mild sensorineural hearing loss in his left ear.

25.7 Outcome

A report was sent to the patient's attorney and an otologic examination was scheduled to obtain a medical evaluation and diagnosis. The audiologist followed-up several weeks later and was informed that the lawsuit had been dropped.

25.8 Key Points

1. When the results from an initial conventional audiological examination reveal discrepancies, the audiologist should not immediately conclude the patient is being uncooperative. The audiologist should first check him or herself, the equipment, and then the patient.
2. If it is determined that a patient is not cooperating for behavioral testing, the first step is to provide the patient the opportunity to change behavior.
3. When pseudohypoacusis is confirmed, behavioral and nonbehavioral audiological procedures are available to document the patient's "true" thresholds. The Stenger test is an excellent procedure when unilateral pseudohypoacusis is suspected because it provides an objective assessment of "true" threshold using the MCIIL procedure.
4. When writing reports on pseudohypoacusic patients rely only on the results from the examination.

Suggested Reading

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26 Ototoxicity and Cisplatin

Steven Smith

A patient with cancer was treated with cisplatin and immediately noticed bilateral tinnitus after receiving treatment. He was referred for an audiological examination and follow-up testing.

26.1 Clinical History and Description

JB is a 39-year-old male who was diagnosed with testicular cancer in mid-June. In early July, he had surgery and the cancer was successfully removed. Because recurrence of testicular cancer is high, JB was treated with cisplatin and gemcitabine to minimize the chance of cancer relapse. The chemotherapy course of treatment consists of receiving three rounds over 3 months of cisplatin along with gemcitabine. In September JB was scheduled to receive his first dose of chemotherapy. Six days after receiving cisplatin and gemcitabine he returned to his oncologist stating that within 24 hours of receiving the drugs he had bilateral tinnitus with the tinnitus in the right ear louder than the tinnitus in the left ear. JB reported that over the next few days the intensity of the bilateral tinnitus decreased, but the tinnitus was still present. He did not report any change in hearing and was referred for an audiological examination.

26.2 Audiological Testing

JB was evaluated with a comprehensive audiological examination including high-frequency audiometry (HFA). Testing revealed normal hearing from 250 to 4000 Hz, sloping to a slight sensorineural hearing loss bilaterally at 6000 and 8000 Hz. Speech recognition thresholds (SRTs) were normal (15 dB HL bilaterally), and word recognition scores (WRSs) were 100% bilaterally. Tympanograms were normal bilaterally. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present bilaterally (500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz), and acoustic reflex decay testing was negative bilaterally at 500 Hz and 1000 Hz. HFA revealed the following hearing thresholds (dB HL) for the right ear: 25 dB at 9000 Hz, 20 dB at 10,000 Hz, 25 dB at 11,200 Hz, 25 dB at 12,500 Hz, 50 dB at 14,000 Hz, and no response (NR) at 16,000 Hz, For the left ear the following hearing thresholds were measured: 25 dB at 9000 Hz, 25 dB at 10,000 Hz, 25 dB at 11,200 Hz, 30 dB at 12,500 Hz, 45 dB at 14,000 Hz, and NR at 16,000 Hz. Table 26.1 summarizes the baseline air conduction thresholds.

26.3 Questions to the Reader

1. What may be the cause of the tinnitus?
2. What audiometric testing should be performed on this patient?
3. How should this patient be followed clinically?
4. What is considered to be a clinically significant change regarding ototoxicity?

Table 26.1 Summary of bilateral baseline audiometric thresholds (dB HL) at 250 Hz to 16,000 Hz.

Frequency (Hz)	Right (dB HL)	Left (dB HL)
250	15	5
500	10	5
1000	15	10
2000	15	15
3000	10	20
4000	15	15
6000	25	25
8000	20	20
9000	25	25
10,000	20	25
11,200	25	25
12,500	25	30
14,000	50	55
16,000	NR	NR

26.4 Discussion of Questions to the Reader

1. What may be the cause of the tinnitus?
   Tinnitus can arise for many different reasons; however, in this case the tinnitus has been caused by cochlear damage due to the administration of cisplatin. It has been reported that hearing loss can be present in 60 to 100%of patients receiving cisplatin. It is challenging, however, to predict the severity of hearing loss that will be caused by cisplatin with any patient as well as how quickly the hearing loss will be noticed by the patient. Some facts are known regarding cisplatin and hearing loss. Cisplatin toxicity usually occurs in the basal (high-frequency) end of the cochlea. As more cisplatin is administered, the more the apical end of the cochlea could begin to be affected. The amount of cisplatin that can cause damage to hearing is not well understood, but it appears that the more cisplatin a patient receives the more hearing loss will be present. It is theorized that cisplatin causes the release of toxic levels of reactive oxygen species, which in turn cause cell death. Research is under way to better understand the interaction between the amount of cisplatin and the resulting hearing loss.
2. What audiometric testing should be performed on this patient?
   A comprehensive audiological examination should be performed on all patients receiving ototoxic drugs. This includes air conduction and bone conduction thresholds, SRTs, WRSs, tympanometry, ARTs, and acoustic reflex decay. In addition, the patient receiving ototoxic medication should undergo HFA. Through the use of HFA a baseline of hearing thresholds at higher frequencies (9000–20,000 Hz) can be measured. This is of great importance because damage to the cochlea

typically begins in the higher frequencies. Therefore, any change in hearing can be observed prior to the hearing loss impacting the speech frequencies and then begin to impact speech recognition in quiet and noise. It is recommended that audiometric testing be performed prior to the patient receiving ototoxic medication so it can then be used as a reference point for future tests. That is, patients can be used as their own control as long as subsequent HFA tests are conducted using the same equipment and test conditions. If changes in hearing thresholds are noted, the physician may be able to change the medication or dosage to prevent further hearing loss. Otoacoustic emissions (OAEs) may also be used as a tool in ototoxic monitoring. Currently, however, there is no guideline as to what represents a significant change in OAEs and what a change in OAEs may mean as it relates to ototoxicity. As already mentioned, ototoxic medication typically initially damages the high-frequency region of the cochlea. Therefore if HFA is not immediately completed prior to administering the ototoxic drug, a decrease in hearing may occur without the audiologist, physician, or patient realizing that the loss in hearing is occurring.

3. How should this patient be followed clinically?
   It is recommended that any patient receiving cisplatin or any ototoxic drug should be given an audiological examination prior to each dose. This should consist of a conventional comprehensive audiological examination along with HFA. The physician ordering the medication should be notified if any significant change occurs in hearing threshold. If significant threshold changes are found, the patient and physician can make a decision as to whether the medication should be continued with hearing compromised or if other treatment options are available. The patient should be advised of any change in hearing and appropriate technology such as hearing aids, hearing assistive technologies (HATs), and cochlear implants may be introduced to assist the patient if any difficulty in hearing should arise. In addition, the audiologist can provide information on communication strategies to the patient as well as the family members/caregivers.
4. What is considered to be a clinically significant change regarding ototoxicity?
   The American-Speech-Language-Hearing Association established what is considered to be a clinically significant change in hearing threshold when ototoxic medication is administered. Significant change is defined as (1) a decrease in hearing threshold of ≥ 20 dB HL at one frequency, (2) a decrease in hearing threshold of ≥ 10 dB HL at two adjacent frequencies, and /or (3) a decrease in hearing thresholds at three adjacent frequencies where responses were previously measured. Any change that is recorded is to be confirmed through a reevaluation.

26.5 Diagnosis and Recommended Treatment

JB was unaware of the possibility that cisplatin could cause tinnitus and hearing loss. He was informed that, because no audiological examination had been performed prior to his initial treatment it would be difficult to determine if any changes in his hearing had occurred. JB stated he was scheduled for his next dose of chemotherapy, which would include cisplatin, within the next week. He was very concerned about this because the tinnitus was bothersome, and he was concerned about having increased difficulty hearing. His physician was contacted with the results of the initial audiological examination, and JB was informed that if he received another dose of cisplatin he should be examined prior to any subsequent dose. JB and his physician determined that the best course of treatment for JB was to continue with the chemotherapy; therefore JB received a second dose of cisplatin.

26.6 Additional Testing

JB returned for an audiological examination 24 hours prior to what would be his third dose of cisplatin. JB stated that after the second dose of cisplatin the tinnitus increased bilaterally within 24 hours, and he does not feel the tinnitus has decreased in loudness. JB also reported that he did not sense there has been any change in his hearing.

JB's hearing thresholds were measured and revealed a slight sensorineural hearing loss at 250 to 500 Hz rising to within normal limits at 1000 to 4000 Hz and then sloping to a mild sensorineural hearing loss in the right ear. Results for the left ear were within normal limits at 250 to 4000 Hz and then sloping to a mild sensorineural hearing loss above 4000 Hz. SRTs and WRSs were not measured due to JB's lack of perceived difficulty with speech and language. HFA revealed the following hearing thresholds (dB HL) for the right ear: 50 dB at 9000 Hz, 50 dB at 10,000 Hz, 50 dB at 11,200 Hz, 60 dB at 12,500 Hz, 60 dB at 14,000 Hz, and NR at 16,000 Hz. For the left ear the following hearing thresholds were measured: 45 dB at 9000 Hz, 50 dB at 10,000 Hz, 50 dB at 11,200 Hz, 55 dB at 12,500 Hz, 55 dB at 14,000 Hz, NR at 16,000 Hz.

Significant changes of 10 dB or greater were noted at 250 Hz, 500 Hz, 8000 Hz, 9000 Hz, 10,000 Hz, 11,200 Hz, 12,500 Hz, and 14,000 Hz for the right ear and 6000 Hz, 8000 Hz, 9000 Hz, 10,000 Hz, 11,200 Hz, 12,500 Hz, and 14,000 Hz for the left ear. A summary of the hearing thresholds and the changes from baseline are summarized in Table 26.2 and Table 26.3.

Table 26.2 Audiometric thresholds (dB HL) at 250 Hz to 16,000 Hz and changes from baseline for the right ear

Frequency (Hz)	Right (dB HL)	Change in right (dB)
250	25	10
500	20	10
1000	10	5
2000	15	0
3000	15	5
4000	15	0
6000	30	5
8000	40	20
9000	50	25
10,000	50	30
12,000	50	25
12,500	60	35
14,000	50	0
16,000	NR	0

Table 26.3 Audiometric thresholds (dB HL) at 250 Hz to 16,000 Hz and changes from baseline for the left ear

Frequency (Hz)	Left (dB HL)	Change in left (dB)
250	10	5
500	5	0
1000	10	0
2000	15	0
3000	20	0
4000	15	0
6000	35	10
8000	40	20
9000	45	20
10,000	50	25
12,000	50	25
12,500	55	25
14,000	55	0
16,000	NR	0

Abbreviation now cited previously in article

JB stated he was unsure if he wanted to receive any additional doses of chemotherapy because he did not want to experience an increase in the loudness of his tinnitus. JB was counseled regarding the decrease in hearing thresholds and that most likely there would be additional decreases in his hearing and an increase in the loudness of his tinnitus if he received additional doses. His physician contacted the audiologist, and it was reconfirmed that if the cisplatin were continued a decrease in hearing would be likely. It was determined that chemotherapy would be provided to the patient without cisplatin, and his hearing thresholds would be reevaluated in a few weeks.

26.7 Outcome

JB returned 3 weeks later stating he had not noticed any change in hearing or tinnitus. JB reported he did not receive the last dose of cisplatin because he did not want his hearing loss or tinnitus to increase. JB states he is becoming more acclimatized to the tinnitus, but that the tinnitus is still bothersome at times. His audiological examination for the right ear revealed a slight hearing loss at 250 to 500 Hz rising to within normal limits at 1000 to 4000 Hz sloping to a mild sensorineural hearing loss. For the left ear, hearing thresholds were within normal limits at 250 to 4000 Hz and then sloping to a mild sensorineural hearing loss above 4000 Hz. Again, SRTs and WRSs were not measured. No significant change was noted from his previous audiological examination. HFA showed no significant changes from the previous examination.

Because JB was concerned with his ongoing tinnitus, he was referred for a tinnitus evaluation. He stated he was becoming less aware of the tinnitus, but he would appreciate more information on tinnitus treatment. JB followed up with an audiologist to obtain information on tinnitus therapy but did not feel he needed any further treatment at the present time.

26.8 Key Points

1. Ototoxicity most likely will occur in the ultra high frequencies and spread to the lower frequencies as more ototoxic medication is administered.
2. Ultra high frequency audiometry is the gold standard with which to monitor patients receiving ototoxic medication.

Suggested Reading

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27 Facial Nerve Monitoring with Electroneuronography

Hillary Snapp

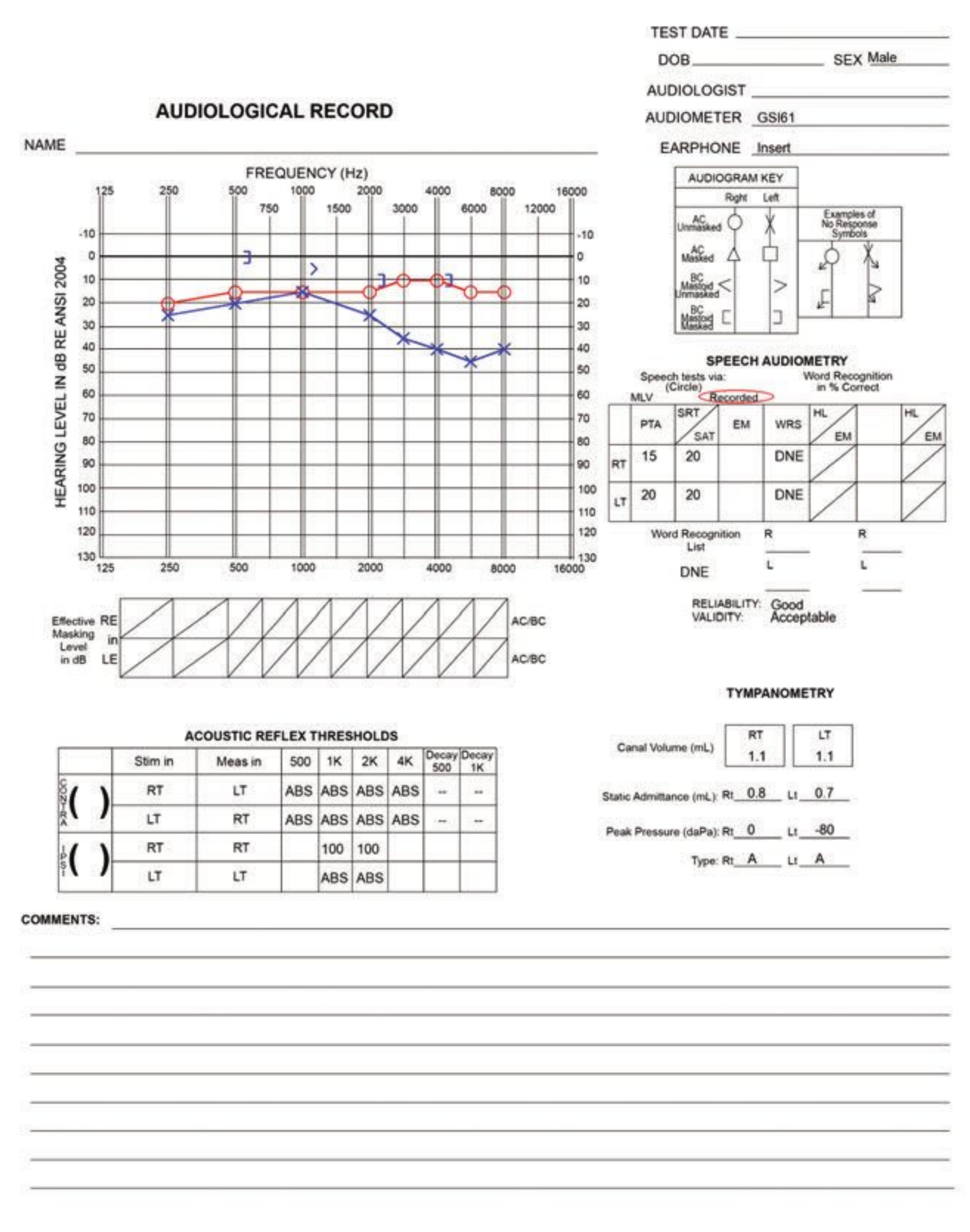
A 21-year-old male presents to the clinic with bilateral facial nerve paralysis.

27.1 Clinical History and Description

JH is a 21-year-old male who sustained severe head injury secondary to a motor vehicle accident. An unrestrained driver, JH presented to the emergency room with bilateral longitudinal temporal bone fractures. A computed tomographic (CT) scan revealed fractures traversing the horizontal segment of the facial nerve canal, small subarachnoid hemorrhage, and pulmonary contusions. JH presented to the neurotology/audiology department 14 days posttrauma and reported a mild decrease in hearing bilaterally and bilateral facial nerve paralysis. There was no evidence of cerebrospinal fluid leakage, otorrhea, or drainage from the ear canals. JH denied otalgia, tinnitus, imbalance, or vertigo but reported a mild tingling sensation in his lips on the right side. Electromyography (EMG) was performed in the hospital, and the patient had been on 60 mg of prednisone daily for 6 days. The neurotologist reported a positive Rinne test bilaterally and House–Brackmann (HB) grade VI/VI. The HB grading system is a widely accepted tool to quantify facial function based on clinical presentation along a six-point scale from normal (HB I) to total paralysis (HB VI). JH was referred for audiological examination and electroneuronography (ENoG).

27.2 Audiological Testing

The audiological examination (Fig. 27.1) revealed that JH had a slight hearing loss at 250 Hz followed by normal hearing at


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols

AUDIOLOGICAL RECORD

NAME

TEST DATE

DOB

SEX Male

AUDIOLOGIST

AUDIOMETER GSI61

EARPHONE Insert

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT	15	20		DNE			
LT	20	20		DNE			

Word Recognition List

DNE

R

L

R

L

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.1	1.1

Static Admittance (mL): Rt 0.8 Lt 0.7

Peak Pressure (daPa): Rt 0 Lt -80

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	ABS	ABS	ABS	--	--
	LT	RT	ABS	ABS	ABS	ABS	--	--
IPSI ( )	RT	RT		100	100			
	LT	LT		ABS	ABS			

COMMENTS:

Fig. 27.1 Results from the audiological examination obtained at the initial presentation 14 days following head trauma resulting in bilateral facial nerve paralysis.

500 to 8000 Hz in the right ear. Masked bone conduction could not be performed in the right ear due to JH's sensitivity to the bone conduction transducer because of the facial and temporal bone fractures. Hearing thresholds for the left ear revealed a slight conductive hearing loss at 250 to 500 Hz rising to normal hearing at 1000 Hz and then gradually falling to a mild conductive hearing loss at 2000 to 8000 Hz with observable air–bone gaps at 500 and 2000 to 4000 Hz. Speech awareness thresholds (SATs) revealed a slight loss and were consistent with the pure-tone average bilaterally. Word recognition scores (WRSs) were not measured. These results are consistent with normal sensorineural reserve bilaterally, suggesting good eighth nerve function. Immittance audiometry revealed normal tympanograms bilaterally, and acoustic reflex thresholds (ARTs) were present at 100 dB HL for 1000 and 2000 Hz ipsilateral stimulation in the right ear, but absent to right and left contralateral stimulation and left ipsilateral stimulation.

27.3 Questions to the Reader

1. Is JH's audiometric configuration consistent with his case history?
2. Why was word recognition not evaluated?
3. Are the results from tympanometry consistent with JH's hearing loss?
4. What does the ART pattern suggest to the audiologist?

27.4 Discussion of Questions to the Reader

1. Is JH's audiometric configuration consistent with his case history?
   Yes, sensorineural and/or conductive hearing loss may occur with trauma to the temporal bone. The type and degree of hearing loss are dependent on the location of the injury. Conductive hearing loss is often due to blood filling the middle ear space (hemotympanum), a cerebrospinal fluid leak, or structural changes to the ossicular chain.
2. Why was word recognition not evaluated?
   The needs of the patient should always be paramount. In a case such as this, obtaining all the audiological data was not as important as obtaining the essential audiological information. In this case, JH is reporting bilateral hearing loss and facial paralysis. The primary goal of the audiologist was to measure hearing and determine the function of the facial nerve. A patient with unilateral facial paralysis often has a difficult time speaking. Consider the limitations for a patient with bilateral facial nerve paralysis. In this case, there were no concerns of impairment of expressive or receptive language or cognitive involvement as a result of the head trauma. In the author's opinion, tympanometry and ARTs provided more important information than assessing word recognition ability. Although JH could not effectively repeat words, he provided a visual signal (i.e., raising his arm) to indicate he was able to detect the speech stimuli, thereby providing the audiologist with an SAT.
3. Are the results from tympanometry consistent with JH's hearing loss?
   If there is a sufficient amount of blood collecting in the middle ear space, the patient may present with a nonmobile tympanic membrane. It is not uncommon, however, for the middle ear air pressure and compliance of the middle ear system to remain intact, despite the presence of conductive hearing loss. This is commonly attributed to disruption of the ossicular chain, resulting in changes to the mechanics of the middle ear system. A complete ossicular dislocation would result in a hypermobile tympanic membrane and middle ear system. The presence of normal middle ear pressure and mobility suggests a partial dislocation without total separation.
4. What does the ART pattern suggest to the audiologist?
   ARTs result in a bilateral contraction of the stapedius muscle in response to a loud sound. The stapedius muscle is innervated by the seventh cranial (facial) nerve. Therefore, in the presence of a facial nerve paralysis, the stapedius muscle is expected to be affected, and the neural pathway must be intact for the stapedius muscle to contract. For facial nerve pathology, a probe effect pattern is expected. That is, the ART will typically be absent when measured on the affected side. Interestingly, JH had ipsilateral ART responses at 100 dB HL on the right side despite no observable facial movement. It is likely that this ART is elevated and reflects the probability that there may be slightly more residual facial nerve function on the right side than on the left side, which is consistent with the ENoG findings.

27.5 Additional Audiological Testing

ENoG is a noninvasive electrophysiological measurement used to examine the integrity of the facial nerve. The nerve is activated using an electrical stimulator placed over the facial nerve. The compound action potential is recorded using surface electrodes placed at the terminal endings of the facial nerve innervation to the muscles of the face. A ground electrode is placed on the patient's forehead.

Injury to the facial nerve will often recover spontaneously. Monitoring facial nerve function by ENoG is used to determine the likelihood of spontaneous recovery. Complete denervation (known as Wallerian degeneration) of the facial nerve occurs over a 72-hour period. For accurate assessment of the damage to the facial nerve, baseline testing should occur after the 72-hour onset of facial paralysis and continue every 3 to 5 days until 21 days post-insult. Electrophysiological responses beyond this point become less reliable, although recovery will classically continue for several months. If facial nerve responses decrease or fail to improve over this period, medical intervention is warranted.

For JH, stimulation of the right facial nerve resulted in a minimal response at 8 ms of 58 µV (□ Fig. 27.2). A masseter response was noted preceding the facial nerve response at 5.3 ms and was verified by several replacements of the stimulator and by little to no observable facial muscle response. Stimulation of the left facial nerve resulted in no observable response. A masseter response was noted at 5.5 ms and was verified as a masseter response by several replacements of the stimulator and by no observable facial muscle response.

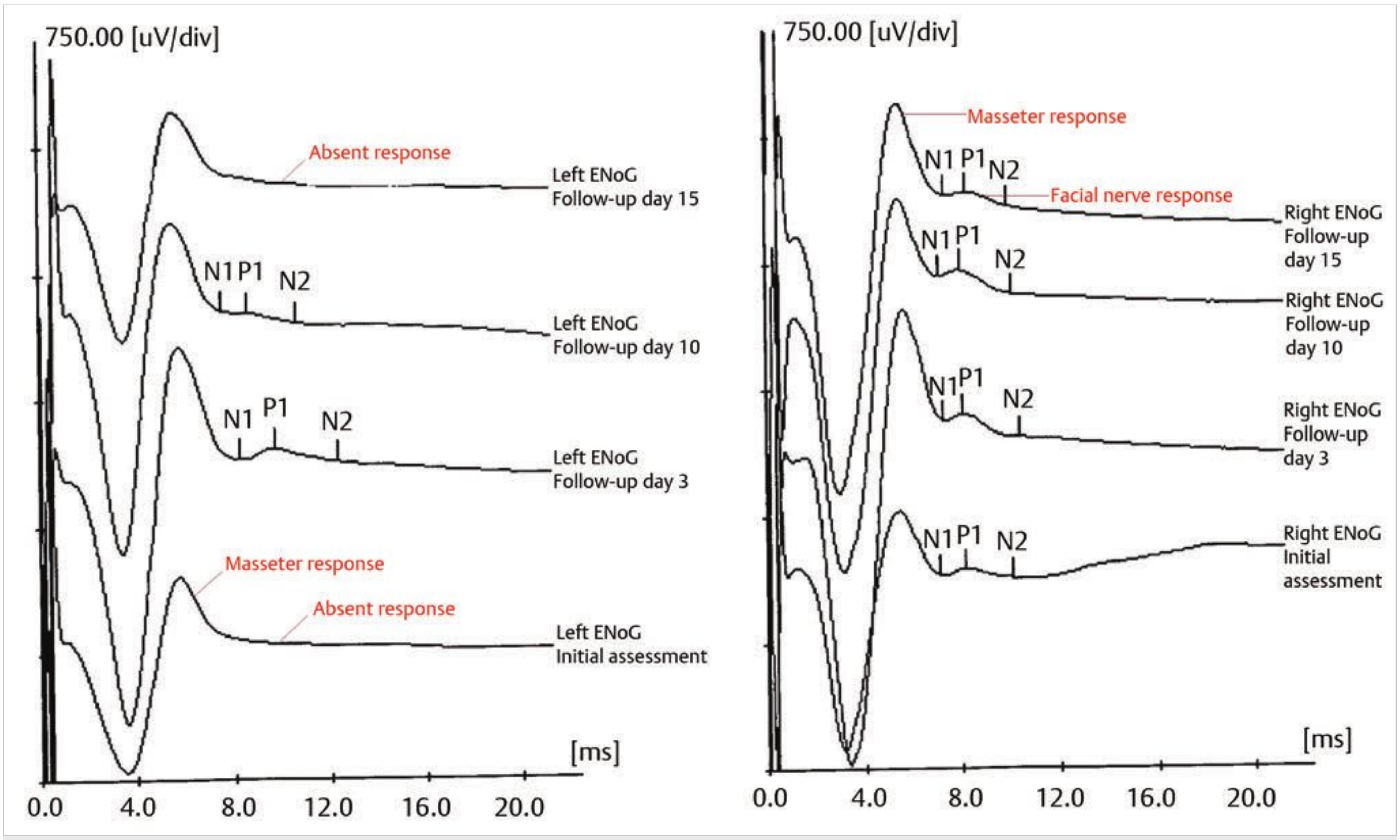


Fig. 27.2 Serial electroneuonographic (ENoG) scans obtained over a 15-day monitoring period, demonstrating initial recovery of the left facial nerve response (as measured by the P_1 to N_2 amplitude) that disappeared at follow-up day 15. In the tracings the right tracing remains intact but does not improve over time. Note that N_1 denotes the initial negative deflection of facial nerve response, P_1 denotes the positive deflection, and N_2 denotes the final negative deflection.

27.6 Additional Questions to the Reader

1. How would an audiologist interpret the findings?
2. Do the ENoG responses reflect what was reported by JH in the case history?
3. What information did the audiologist obtain from facial nerve monitoring?
4. What is the appropriate time frame for completing facial nerve monitoring in a case of facial paralysis?

27.7 Discussion of Additional Questions to the Reader

1. How would an audiologist interpret the findings?
   Facial nerve function is assessed by using electrical stimulation at the stylomastoid foramen to trigger a compound action potential that is measured by surface electrodes on the face (Fig. 27.3). The surface electrodes are placed along the nasolabial fold at the base of the nose and the corner of the mouth because this represents the most peripheral points of facial nerve innervation for the musculature of the upper and lower regions of the face. The facial nerve response will typically occur at around 8 ms and is characterized by an initial negative deflection (N_1), followed by a large positive deflection (P_1), followed by a final negative deflection (N_2). The amplitude of P_1 to N_2 is the primary measurement used to assess function of the facial nerve. Generally, ENoG interpretation is a calculation of percent of denervation based on a comparison of the response amplitude in µV for the involved side versus the uninvolved side.

$$\%\ \text{Denervation} = \frac{100 - \text{Amplitude of Involved Side}}{\text{Amplitude of Uninvolved Side} \times 100} \qquad (27.1)$$

   Typically, a denervation of less than 90% is an indication to continue to watch and wait for spontaneous recovery, whereas greater than 90% denervation is an indication for surgical intervention. Thus interpretation is more challenging for cases of bilateral involvement because the percent denervation cannot be calculated. Latency does not carry much diagnostic value, although it is important for accurate identification of P_1. Note the large positive deflections at 5 msec. This large response is from the masseter muscle and should not be confused as a response from the facial nerve. Due to its proximity to the stimulator site, the masseter muscle will generate a response that can be seen preceding the facial nerve response. Altering the position of the stimulator (Fig. 27.3) anteriorly to intentionally stimulate the masseter muscle can be used as a cross check by comparing the latency of the response.
2. Do the ENoG responses reflect what was reported by JH in the case history?
   Yes. A normally functioning facial nerve will trigger a response from 500 to 2000 µV or more. A cursory view of the

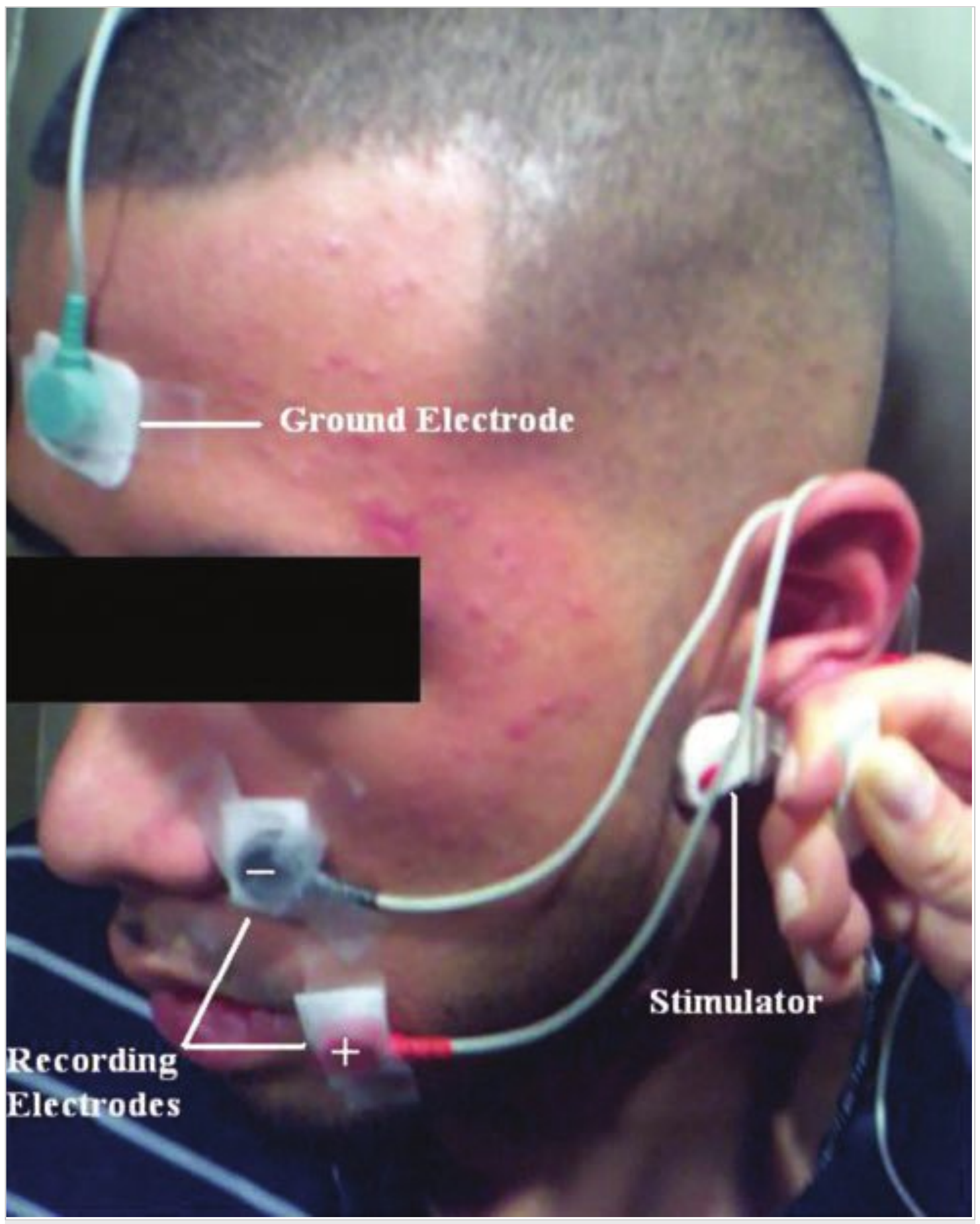

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Fig. 27.3 Placement for the stimulating and recording electrodes used in electroneuronographic measurement. Stimulating electrodes are placed inferior to the earlobe with the negative electrode posteriorly located and the positive electrode anteriorly located to the earlobe. Careful placement of the stimulator is essential for optimal stimulation of the facial nerve responses because even a slight misplacement anteriorly will result in a primary masseter response. The recording electrodes are placed along the nasolabial fold.

tracings (Fig. 27.2) revealed that on the first visit the response on the left is absent and the amplitude of the response on the right side is very low (58 µV).As already noted, ENoG amplitudes range from 500 µV to greater than 2000 µV. This finding is consistent with JH's reported bilateral involvement and suggests absence of facial nerve function on the left side and minimal residual function on the right side. In cases where both right and left facial nerves are affected, a denervation calculation is not appropriate. For JH, the approach was to monitor the changes in the response amplitude for each side independently (initial assessment, 3, 10, and 15 days post initial assessment). A decrease in amplitude suggests further distress of the facial nerve, whereas an increase in amplitude over time suggests facial nerve recovery.

3. What information did the audiologist obtain from facial nerve monitoring?
   The initial overall response amplitude of 58 µV was low (normal = ≥500 µV) for the right side and essentially absent on the left side. Follow-up ENoGs revealed a stable response on the right side and suggested facial nerve recovery on the left side. Over the next several days, however, the left response began to decline and eventually disappear completely. This decline in amplitude suggested to the audiologist that the left nerve was in critical distress and required surgical intervention. The patient was scheduled for surgical decompression of the left facial nerve. Decompression may be performed to reduce swelling and/or relieve pressure on the facial nerve by removing a small portion of the temporal bone.
4. What is the appropriate time frame for completing facial nerve monitoring in a case of facial paralysis?
   Facial nerve monitoring is used when there is concern that greater than 90% denervation has or will occur. With less than 90% denervation, there is a high rate of spontaneous recovery. Wallerian degeneration (denervation of the facial nerve) takes approximately 72 hours to occur and is typically monitored to 21 days postonset for consideration of surgical intervention. JH's case is unique in that JH was monitored for 29 days. Monitoring continued beyond the time frame because JH's nerve initially appeared to be recovering (Fig. 27.2) without intervention.

27.8 Diagnosis and Recommended Treatment

JH was diagnosed by the neurotologist with bilateral facial nerve paralysis with a VI/VI HB scale. On the HB scale, a grade 1 presentation represents normal function, grade 2 indicates mild weakness, grade 3 moderate weakness with good (or normal) eye closure, grade 4 moderate weakness with no volitional eye closure, grade 5 severe weakness, and grade 6 total facial paralysis. At the 10-day follow-up, JH reported he had headaches when he woke in the morning and also felt decreased sensation in his lips when he lay down. JH had to sleep on his back due to the multiple injuries related to his accident. It was determined that the bilateral fractures were causing the base of JH's skull to shift slightly during sleep, resulting in increased risk to the left facial nerve. The ENoG results confirmed that facial nerve responses were decreasing on the left side, and JH was scheduled for left facial nerve decompression.

27.9 Outcome

At the 1-week postoperative visit, JH presented with upper brow and lip movement on the left side of his face. By 2 months JH presented with significant facial recovery (III/VI HB). JH returned for final follow-up 6 months after the decompression surgery and presented with full facial recovery (1/6 HB), bilaterally. The conductive hearing loss in the left ear persisted, consistent with disruption of the ossicular chain. Although JH complained of left tinnitus, decreased hearing, and aural fullness in the left ear, he declined medical management due to the mild degree of hearing loss. Tympanometry remained normal and ARTs did not recover.

This case report represents a unique presentation of bilateral facial nerve paralysis and highlights the value of ENoG as a monitoring tool for consideration of surgery in conservative medical management of such cases.

27.10 Key Points

1. ENoG is an objective method for evaluating facial nerve function. For many patients with facial nerve paralysis,

monitoring the facial nerve through ENoG is essential for determining course of treatment and prognosis for recovery. Clinical assessment of this function will often determine the need for surgical intervention.

2. In general, facial nerve monitoring is used when there is concern that greater than 90%denervation has or will occur. With less than 90%denervation, there is a high rate of spontaneous recovery.
3. In cases of bilateral involvement, a denervation calculation is not appropriate. Rather, it is more appropriate to review overall amplitude of the response and closely monitor for changes in facial nerve function to determine the need for surgical intervention.
4. Patients with temporal bone fracture are also at risk for conductive, mixed, or sensorineural hearing loss. Comprehensive evaluation of auditory function is an important component of the diagnostic evaluation of these patients. Several limitations to testing may be associated with patients presenting with head trauma. This may require the clinician to identify alternate means of assessment in order to provide a complete evaluation when challenged by these limitations. Modifications to the test battery should always be based on assessment of the patient's needs and ensure that the accuracy or validity of the testing has not been negatively impacted.

Suggested Reading

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28 Acquired Auditory Neuropathy in Human Immunodeficiency Virus/Acquired Immunodeficiency Syndrome

Brad A. Stach and Virginia Ramachandran

A 49-year-old male presented with sudden hearing disorder in his right ear that he associated with a lumbar puncture.

28.1 Clinical History and Description

GS was 49 years old at the time of his initial clinical encounter. He reported a sudden hearing loss in his right ear that he associated with a lumbar puncture. He also reported long-standing profound hearing loss in his left ear, which he remembers having in childhood. At this initial encounter, GS reported pursuing assessment of his hearing in the hope of receiving additional social-service disability income.

GS was a patient known to our hospital for 15 months prior to his initial audiological examination. His first encounter with the hospital was in the emergency room (ER), where he sought services for difficulty breathing, nausea, vomiting, and abdominal pain. He was undergoing treatment from another facility for pneumonia. Examination at the time confirmed the presence of pneumonia and also revealed significant hypertension. His nausea and vomiting resolved, and he was supposed to have been given intravenous antibiotics and discharged. Prior to discharge, however, he complained of shortness of breath, became diaphoretic, and began hyperventilating. He was admitted to the hospital for management of his pneumonia with sepsis. Serology studies were positive for hepatitis C antibody, hepatitis B core total antibody, and human immunodeficiency virus (HIV). He was discharged from the hospital 4 days later and placed on antiretroviral medication.

GS returned to the ER 8 months later, presenting with headache, vomiting, and a rash on his back. He was in significant distress and was admitted immediately with elevated blood pressure, heart rate, and fever. After his vital signs were controlled, he underwent a diagnostic lumbar spinal puncture. Analysis of his cerebrospinal fluid (CSF) was positive for cryptococcal meningitis. He was placed on intravenous medication and began to improve clinically. GS was discharged 2 weeks later and placed on oral antifungal medication.

GS's next visit to the ER was 4 months later when he presented with a 3-day history of headache, fever, photophobia, generalized weakness, nausea, and vomiting. A lumbar puncture was performed in the ER, which demonstrated elevated CSF pressure. A urine drug screen was positive for opioids, cocaine, and antidepressants. He was admitted to the infectious disease floor and placed on intravenous (IV) antibiotic and antifungal medications. A second lumbar puncture revealed reduced CSF pressure and significant positive signs of cryptococcal infection. By day 3 of therapy, GS was significantly improved subjectively. After 7 days of therapy, another lumbar puncture was performed and showed a significant reduction in CSF cryptococcal antigen. GS was discharged after 11 days of therapy. His discharge diagnoses included malignant hypertensive heart disease and HIV-related infection.

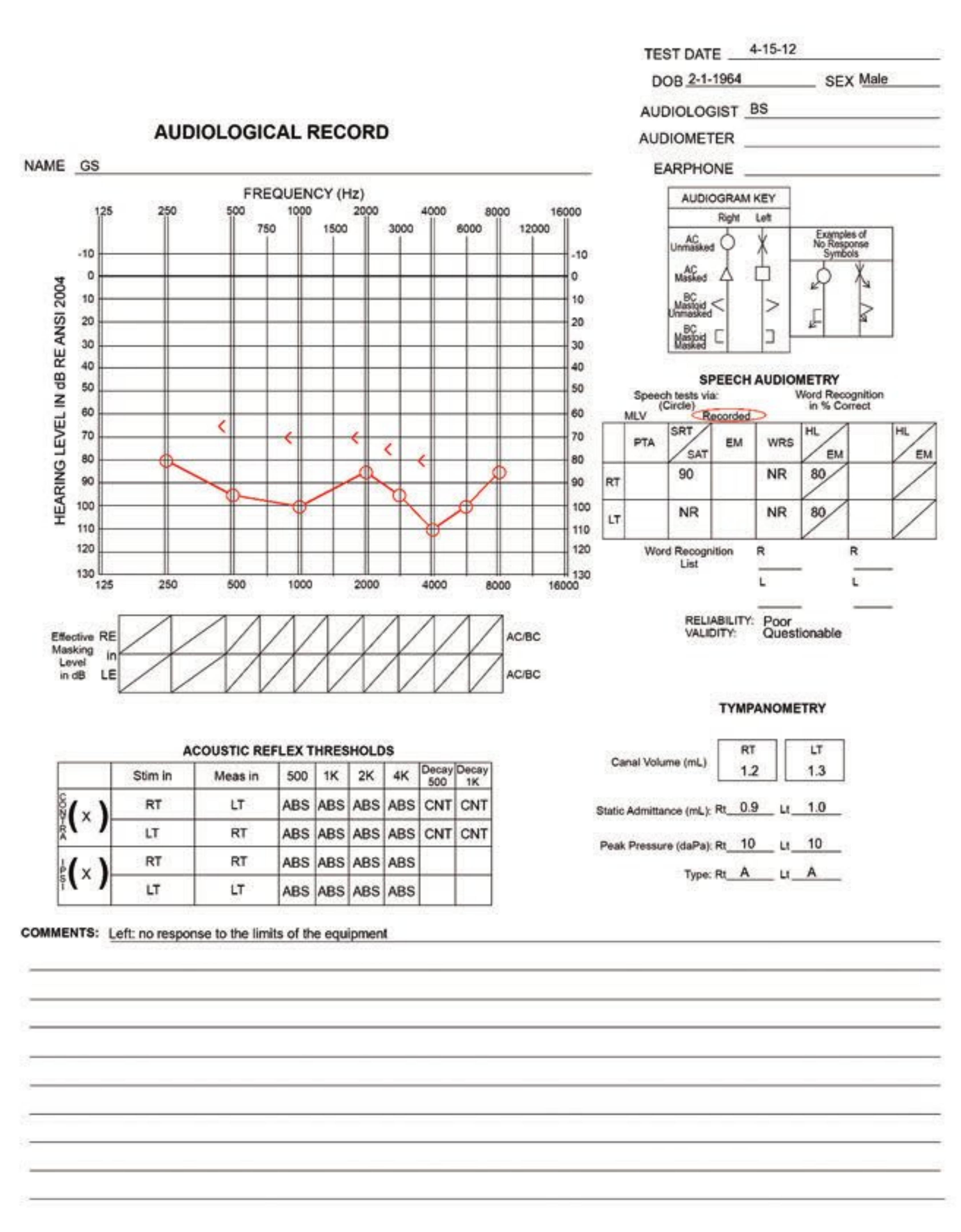
GS reported that his hearing loss occurred during his next hospital admission 3 months later. This admission was precipitated by a visit to the ER following 4 days of headache, blurred vision, nausea, and vomiting. GS was once again admitted to the infectious disease floor and placed on multiple IV medications. By this time, GS was receiving serial lumbar punctures daily. Following one procedure, GS reported the onset of right-ear hearing loss and was referred for an audiological examination. A note was made in the medical record that the antifungal drug, amphotericin B, was discontinued following the patient complaint due to the potential for auditory neurotoxicity.

28.2 Audiological Testing

GS was an inpatient at the time of his initial audiological examination, escorted to the service in a wheelchair with active IV lines. GS appeared lucid throughout the case history and strained only slightly when interacting auditorily. GS reported no hearing in his left ear and that he had begun having trouble understanding speech in his right ear. GS associated the onset of the right-ear hearing loss with one of the lumbar punctures and felt that his hearing had been getting worse while he was in the hospital. GS's case history was negative for tinnitus, fullness, and vertigo. He reported no history of ear infections or other otologic disease. Otoscopy revealed normal ear-canal anatomy, and tympanic membranes were clearly visible and appeared normal.

Results of the first attempt at pure-tone audiometry are reported in Fig. 28.1. The patient did not respond to speech or pure-tones by air or bone conduction in the left ear at equipment limits. Results for the right ear revealed speech awareness thresholds (SATs) of 90 dB HL and pure-tone air conduction thresholds in the severe to profound range across the frequencies. Bone-conduction responses were noted to be vibrotactile at 65 and 70 dB HL. The patient did not appear to be able to recognize any of the words presented for word recognition testing.

Immittance audiometry revealed type A tympanograms bilaterally with normal static admittance, middle-ear pressure, and ear-canal volume bilaterally. All crossed (i.e., contralateral) acoustic reflex thresholds (ARTs) and uncrossed (ipsilateral) ARTs were absent bilaterally. The absence of all ARTs with sound presented to the left ear (i.e., left contralateral and left ipsilateral) was consistent with the patient's report of left-ear deafness from childhood. The absence of all ARTs with sound presented to the right ear (i.e., right contralateral and right ipsilateral) is suggestive of a disorder of the right afferent portion of the reflex arc and consistent with an organic disorder relating to the patient's complaint of a newly acquired hearing loss on the right ear.



AUDIOLOGICAL RECORD

NAME GS

TEST DATE 4-15-12

DOB 2-1-1964 SEX Male

AUDIOLOGIST BS

AUDIOMETER

EARPHONE

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		90		NR	80		
LT		NR		NR	80		

Word Recognition List R L R L

RELIABILITY: Poor
VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.3

Static Admittance (mL): Rt 0.9 Lt 1.0

Peak Pressure (daPa): Rt 10 Lt 10

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS: Left: no response to the limits of the equipment

Fig. 28.1 Results of initial pure-tone and speech audiometry for GS.

Because of the discrepancy between the audiometric outcomes and the patient's communication function during the case-history interview, a second audiologist was enlisted to assist in determining the organicity of the disorder and to establish valid hearing-sensitivity thresholds.

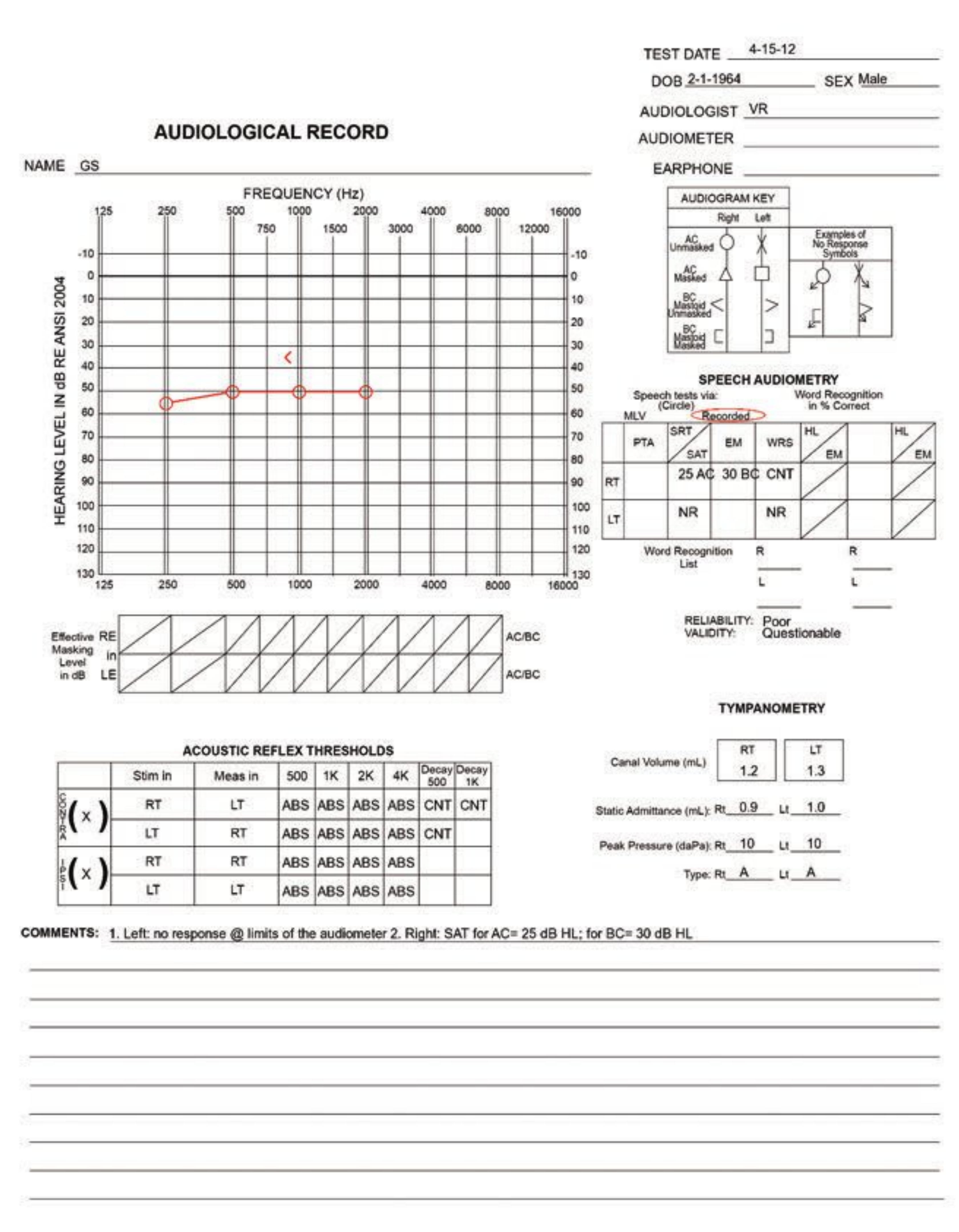
The patient was reinstructed on expected behaviors for measures of pure-tone and speech audiometry. Results of the second attempt at pure-tone audiometry are reported in Fig. 28.2. Once again, the patient did not respond to speech or pure tones by air or bone conduction in the left ear at equipment limits. Responses in the right ear, however, improved following the reinstruction. Right-ear results revealed SATs of 25 dB HL by air conduction and 30 dB HL by bone conduction. Pure-tone air-conduction thresholds were approximately 50 dB HL at the frequencies tested, although responses were noted to be very inconsistent and of poor reliability. A bone-conduction response was noted at 35 dB HL at 1000 Hz. The patient's behavior during speech audiometric testing was also noted to be inconsistent. During speech threshold testing, the patient responded with half-word spondees, repeated the words at times, and "could not understand" at other times.

Immittance measures were repeated and revealed similar to the initial findings. Tympanograms, static immittance, ear canal volumes, and middle ear pressure were all normal bilaterally. All ARTs were absent bilaterally.

The absence of ARTs with sound presented to the right ear suggested the presence of an organic disorder. The patient's behavior during testing suggested malingering, or at least an exaggeration of an organic disorder.

Distortion-product otoacoustic emission (DPOAE) measures were then completed. Results are reported in Fig. 28.3. Not unexpectedly, DPOAEs were absent on the left ear. Results on the right ear, however, showed fairly robust emission amplitudes across the frequency range tested. These results are consistent with normal cochlear outer hair cell function.

The overall pattern of results in the left ear is consistent with a profound sensorineural hearing loss, including consistent absence of behavioral responses, SATs matching the audiometric thresholds, absent ARTs with stimulation to the left ear, and absent DPOAEs. The overall pattern of results in the right ear is consistent with exaggerated or feigned hearing loss, including inconsistent mild to moderate sensitivity loss, normal


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols

AUDIOLOGICAL RECORD

NAME GS

TEST DATE 4-15-12
DOB 2-1-1964 SEX Male
AUDIOLOGIST VR
AUDIOMETER
EARPHONE

AUDIOGRAM KEY

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT		25 AC	30 BC	CNT			
LT		NR		NR			

Word Recognition List R L R L

RELIABILITY: Poor
VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.3

Static Admittance (mL): Rt 0.9 Lt 1.0
Peak Pressure (daPa): Rt 10 Lt 10
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS: 1. Left: no response @ limits of the audiometer 2. Right: SAT for AC= 25 dB HL; for BC= 30 dB HL

Fig. 28.2 Pure-tone and speech audiometry results for GS following reinstruction.

DPOAEs, SATs that do not match the audiometric thresholds, and communication function that does not match the degree of admitted hearing loss. GS was referred for audiological reexamination and evoked potential audiometry in an effort to estimate hearing sensitivity in the right ear.

28.3 Questions to the Reader

1. This patient has HIV infection that appears to be fairly advanced. Is HIV/AIDS associated with hearing disorder?
2. How do you reconcile the absence of ARTs with sound presented to the right ear in the presence of normal or nearly normal OAE amplitudes?
3. Could GS's "functional" behavior be mimicking an auditory processing disorder?

28.4 Discussion of Questions to the Reader

1. This patient has HIV infection that appears to be fairly advanced. Is HIV/acquired immunodeficiency syndrome (AIDS) associated with hearing disorder?
   Yes, auditory disorder is quite prevalent among adults and children with HIV infections. Prevalence rates of auditory disorder in adults range from 20 to 50% The causes of hearing disorder relating to HIV/AIDS are manifold and can affect the cochlea and the peripheral and central auditory nervous system. Cochlear disorder can result from opportunistic infections, such as otosyphilis or meningitis, occurring as a result of the immunodeficiency caused by HIV. Cochlear disorder can also be caused by ototoxicity relating to the antire-

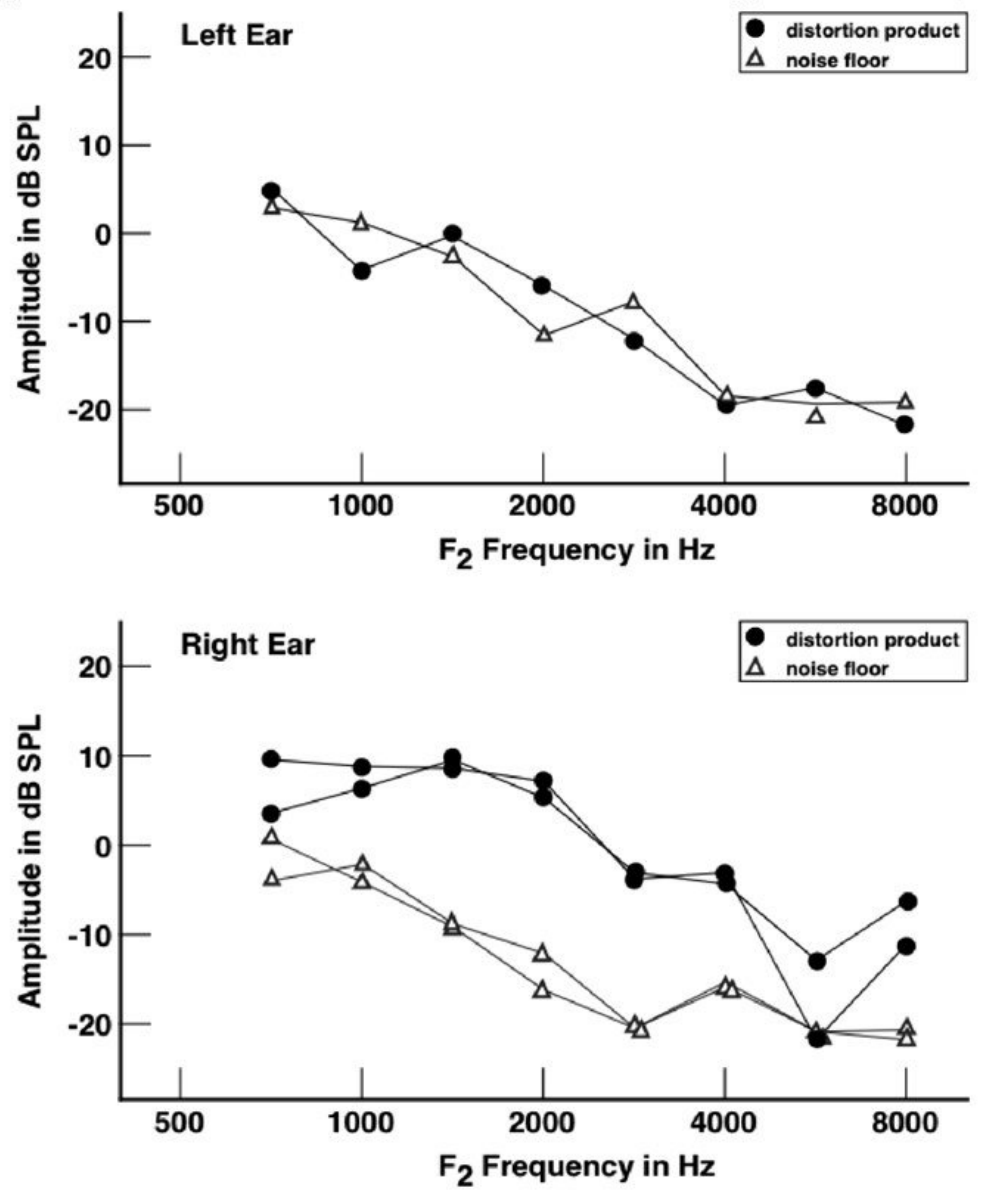


Fig. 28.3 Distortion-product otoacoustic emission amplitude as a function of F2 frequency for GS. Right-ear responses were replicated.

troviral treatment of the HIV and also the antibiotic, antiviral, and antifungal treatments of opportunistic infections. Peripheral and central nervous system disorders are also quite common and usually occur as the direct effect of HIV infection on nervous-system tissue. Changes to the auditory nervous system can result in hearing-sensitivity loss, although the disorder is far more commonly revealed by abnormalities in auditory evoked potentials.

2. How do you reconcile the absence of ARTs with sound presented to the right ear in the presence of normal or nearly normal OAE amplitudes?
   It is difficult to reconcile when considering the most common types of hearing loss. Results from the left ear are consistent: a profound hearing sensitivity loss, absent ARTs with stimuli presented to the left ear for contralateral and ipsilateral presentation, and absent DPOAEs all corroborated a sensory disorder of the left cochlea. Results from the right ear are not. Robust DPOAEs in an ear with behavioral evidence of no more than a mild sensitivity loss would argue for the presence of ARTs with sound presented to the right ear. Their absence in the presence of normal DPOAEs suggests a complexity of the disorder that might not be so easily explained as "functional."
3. Could GS's "functional" behavior be mimicking an auditory processing disorder?
   Most auditory processing disorders are too subtle in their effect to influence a patient's ability to detect pure tones. Even in patients with clear deficits in speech recognition, pure-tone thresholds are easy to obtain and often normal. In some extreme cases, however, such as in patients with bilateral temporal lobe lesions, there appears to be what has been described as "cortical deafness," in which pure tones are apparently not recognized. It is reasonable to assume that some patients will fall between these two extremes and present with sufficient difficulty in listening for pure tones that their behavior takes on the inconsistent appearance of those who are feigning or exaggerating hearing loss.

28.5 Additional Audiological Testing

GS returned for audiological reexamination and evoked-potential audiometry 2 days later. Once again, no behavioral responses were obtained to speech or pure-tone stimuli in the

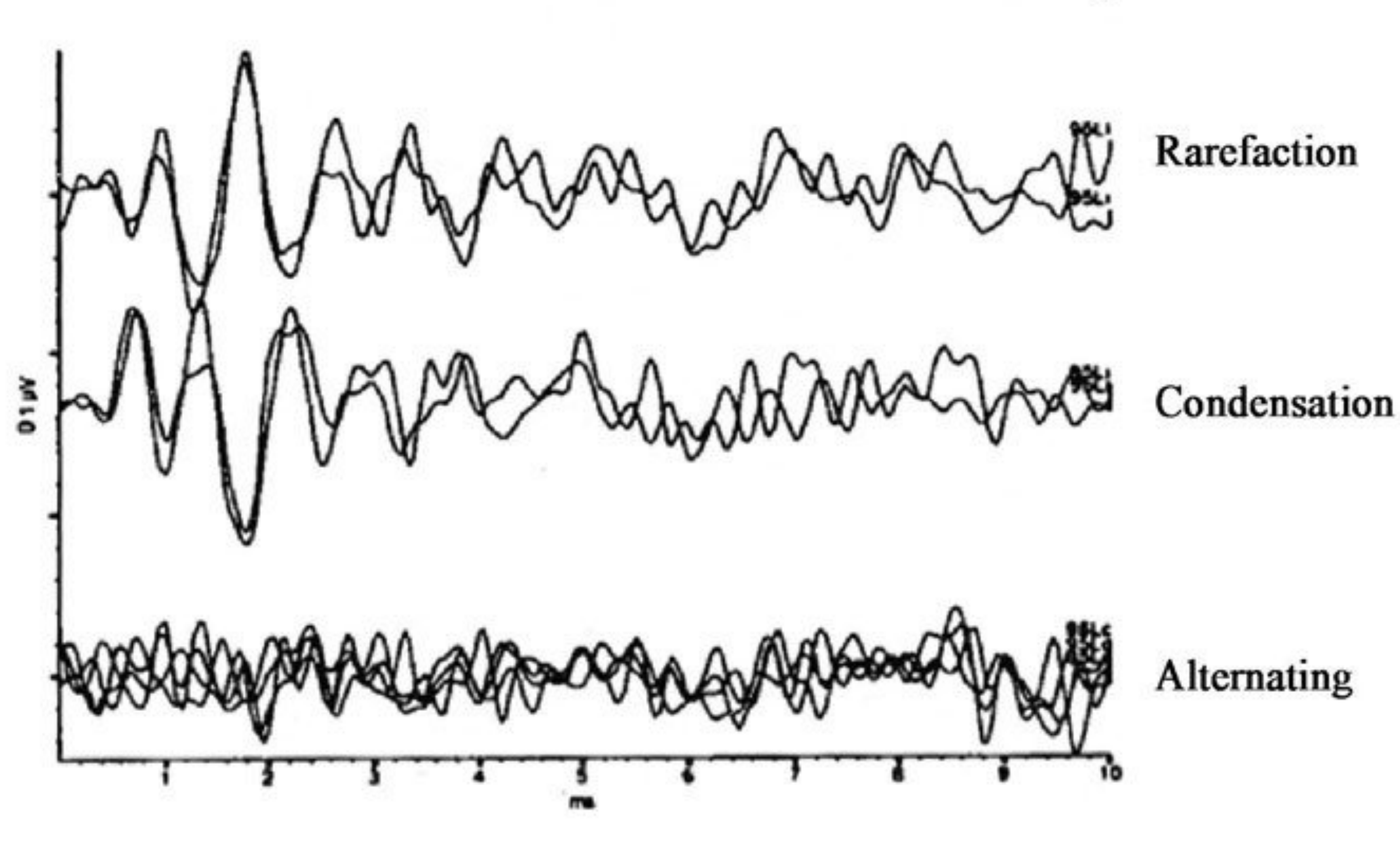


Fig. 28.4 Auditory brainstem response from GS. Responses are from the right ear to rarefaction, condensation, and alternating click stimuli presented at 95 dB nHL at a rate of 21.1/s.

left ear. A SAT of 25 dB HL was obtained in the right ear. Although GS responded to some of the pure tones, reliable and valid thresholds could not be established in that ear.

Auditory brainstem response (ABR) testing was completed to estimate hearing sensitivity. ABRs were absent to click stimuli of alternating, condensation, and rarefaction polarity presented to the left ear at equipment limits. These results are consistent with the long-standing sensorineural hearing loss on the left.

ABR results in the right ear were quite revealing. No synchronous responses were observed to alternating polarity click stimuli presented at 95 dB nHL. When single-polarity stimuli were used, repeatable cochlear microphonics were recorded that changed phase with click-polarity change. Results are shown in Fig. 28.4. No synchronous ABR waves were observed beyond the cochlear microphonic.

Auditory steady-state response (ASSR) measures were absent at all frequencies tested in both ears at equipment limits across the audiometric frequencies. This is consistent with the sensorineural hearing loss in the left ear and the lack of synchronous neural response on the right.

Late-latency responses (LLR) to pure tones were also measured. In the left ear, no responses were noted at equipment limits at 500 or 2000 Hz. In the right ear, responses were observed down to 15 dB at 500 Hz and 20 dB at 2000 Hz.

The audiological diagnosis in the left ear did not change based on evoked-potential audiometry. The overall pattern of results remained consistent with a profound sensorineural hearing loss, including consistent absence of behavioral responses, SATs that match the audiogram, absent ARTs with sound to the left ear, absent DPOAEs, and absent ABR, ASSR, and LLR.

The audiological diagnosis in the right ear changed considerably with the discovery of a lack of synchrony of auditory nerve function. The overall pattern of results in the right ear is consistent with auditory neuropathy, including inconsistent responses to pure tones, normal DPOAEs, absent ARTs, poor speech recognition, and preserved cochlear microphonics, absent ABR and ASSR, and present LLRs.

28.6 Diagnosis and Recommended Treatment

Prior to discharge from the hospital, transaxial computed tomographic (CT) images of the brain were obtained from the skull base to the vertex. Results showed marked periventricular white matter disease, as well as white matter disease in the pons and the middle cerebral peduncle. Based on these results, the patient was diagnosed with HIV encephalopathy.

The patient was eventually discharged from the hospital and did not pursue audiological treatment options. Four months after discharge, he was readmitted to the hospital with headache, neck and back pain, and acute hypertension. His intracranial pressure had worsened, as had his meningitis. Staging of his disease progress was elevated from HIV-related disease to terminal AIDS. He was discharged under hospice care and died shortly thereafter.

28.7 Additional Questions to the Reader

1. Can a lumbar puncture cause hearing loss?
2. Can HIV/AIDS cause auditory neuropathy?
3. Were GS's behavioral audiometric outcomes real, or was he exaggerating?

28.8 Discussion of Additional Questions to the Reader

1. Can a lumbar puncture cause hearing loss?
   Yes. Hearing loss following lumbar puncture has been reported, although it appears to be rare and most often transient. In cases of permanent changes, the expectation would be for the DPOAEs to be absent, reflecting changes in overall cochlear function as a result. Is it possible that GS

experienced a temporary change in his hearing following lumbar puncture that served to direct his attention to the speech recognition problems relating to his auditory neuropathy? That would be pure speculation, but it seems a more likely explanation than that the lumbar puncture would somehow result in auditory neuropathy.

2. Can HIV/AIDS cause auditory neuropathy?
   Yes. A recent report suggests that auditory neuropathy can occur as a result of cryptococcal central nervous system infection in immunocompromised patients.
3. Were GS's behavioral audiometric outcomes real, or was he exaggerating?
   We will never know. GS stated during the case-history interview that he was interested in increased disability funding relating to the apparently new problem he was having with his hearing. Some of his behaviors during the audiological examination were classically functional. Yet from all that clinicians are learning about auditory neuropathy, it is not difficult to imagine that the disorder might result in difficulty listening for pure tones and speech recognition poor enough that responses might appear to be functional.

28.9 Key Points

1. Patients with compromised immune systems can have auditory disorders from multiple causes relating to the disease that compromised the immune system, opportunistic infections that result because of compromised immunity, or ototoxicity from the medications used to treat the primary disease and/or the opportunistic infections. Auditory disorder can occur in the cochlea as well as the peripheral and central auditory nervous systems.
2. The authors present a case in which a patient with HIV disease, hypertension, and cryptococcal meningitis developed a hearing disorder that he related to a lumbar puncture procedure.
3. Clinical outcomes in this patient appear to show auditory neuropathy as the underlying cause of the auditory complaints.

Suggested Reading

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29 When Is a Difference in Word Recognition Scores Significant?

Michael Valente and L. Maureen Valente

One goal of the audiological examination is to provide the patient and other healthcare providers with accurate audiological results, interpretation, and appropriate follow-up recommendations. Often, patients are seen more than once for hearing healthcare. Examples of services performed include ototoxic monitoring, annual evaluations, and initial and postoperative otologic care such as surgery and delivery of medication. One key audiological finding that may suggest a significant change, or lack of change, in hearing is the word recognition score (WRS). Often, clinicians apply an arbitrary indicator (i.e., a “change of 10%”) when reporting that a change in WRS is significant. This case report provides information on how to use critical differences between WRS as a means to make better decisions about the change or lack of change in WRS between examinations.

29.1 Clinical History and Description

This case report describes an 85-year-old male with long-standing bilateral asymmetric sensorineural hearing loss. Visits have been scheduled annually as standard follow-up care to determine if changes in hearing may have required programming changes to his hearing aids that were fit in 2007. This case report highlights differences in hearing between an audiometric examination obtained in 2012 compared with results obtained in 2011. At each evaluation, the patient did not report tinnitus in either ear, recruitment, otalgia, dizziness, fullness, vertigo, or excessive exposure to noise. The patient did, however report increased difficulty recognizing speech in noise or female talkers, and difficulty communicating on the telephone.

After the audiometric examination in 2011, the patient underwent Mohs micrographic surgery for basal cell carcinoma of the right concha. After surgery, the patient reported fullness in his right ear and observation by his otolaryngologist revealed that the external meatus narrowed with a prolapsing portion of the conchal cartilage at the superior aspect of the meatus. In addition, the inferior portion of the cartilage had been resected in the past for reasons unknown to the authors. With gentle manipulation of the auricle by his physician, the prolapsing segment of the cartilage could be pulled up and away, but the patient was unable to discern if this maneuver alleviated the sensation of fullness. With respect to hearing healthcare, the superior conchal cartilage prolapse limited the patient’s ability to fully insert his right hearing aid. The patient was counseled by his physician that conchal cartilage surgical revision could be performed to try to correct this, but the patient decided against this. Due to his continuing swollen ear canal on the right side, the patient was no longer able to use his custom earmold, and his right hearing aid was coupled to his ear canal using a thin tube with a very small dome.

29.2 Audiological Testing

The patient was initially seen in 2007. An audiometric examination at that time revealed hearing to be within normal limits bilaterally from 250 to 2000 Hz, followed by a moderate to moderate severe sensorineural hearing loss (SNHL) in the right ear and a mild to moderate severe SNHL in the left ear at 3000 to 8000 Hz. Speech Recognition Thresholds (SRTs) were within normal limits bilaterally, and WRSs revealed slight difficulty in the right ear and normal ability in the left ear. WRSs were completed at the patient’s Most Intelligible Level (MIL) using a recorded Northwestern University Auditory Test No. 6 (NU-6) word list with a female talker. Tympanometry and acoustic reflex thresholds to contralateral and ipsilateral stimulation at 500 to 4000 Hz were within normal limits. The patient obtained medical clearance for amplification and returned for a hearing aid evaluation (HAE). The patient was eventually fit with bilateral hearing aids with a thin tube and custom open earmolds. From that point, the patient’s hearing was monitored annually (□ Table 29.1) where hearing thresholds, SRT, and WRS were essentially unchanged until his most recent audiometric examination in 2012.

The results from the most recent audiological evaluation were unremarkable with the exception of a 10 dB decrease in hearing thresholds in the right ear at 250 and 3000 to 4000 Hz. In addition, WRSs revealed a decrease of 40% in the right ear and 12% in the left ear. A steady but small decline in the WRSs

Table 29.1 Summary of air conduction hearing thresholds (dB HL) at 250 to 8000 Hz, Speech Recognition Threshold (SRT in dB HL), and Word Recognition Scores (WRSs in %) for the right and left ears from 2007 to 2012.

Right Ear									Left Ear										
Date	250	500	1,000	2,000	3,000	4,000	6,000	8,000	WRS	250	500	1,000	2,000	3,000	4,000	6,000	8,000	SRT	WRS
3/07	15	5	0	20	60	65	70	15	88%	15	10	10	15	35	60	65	70	10	98%
3/08	15	10	5	20	55	65	65	15	88%	15	15	10	20	35	55	70	70	10	98%
3/09	15	10	10	20	55	60	65	15	92%	15	15	10	20	35	55	60	75	10	92%
3/10	10	5	10	20	55	60	75	15	88%	15	10	10	15	35	60	65	75	10	88%
3/11	10	10	15	35	60	75	70	20	80%	15	10	10	15	35	60	65	65	15	82%
3/12	25	5	20	40	70	75	70	25	40%	20	15	10	20	35	55	75	70	15	70%

from 98% in 2007 to 70% in 2012 was noted, whereas the WRS in the right ear was essentially stable until the most recent examination in 2012. At the time of the examination in 2012, tympanometry revealed normal pressure, ear canal volume, and peak admittance bilaterally. Acoustic reflex thresholds to contralateral stimulation were absent or elevated bilaterally. Acoustic reflex thresholds to ipsilateral stimulation were absent for the right ear at 500 to 4000 Hz. They were within normal limits at 500, 2000, and 4000 Hz but absent at 2000 Hz for the left ear. Acoustic reflex decay was abnormal in the right ear at 1000 Hz. A letter, summary report, and copy of the audiometric evaluation were faxed to the patient's primary care physician recommending a referral to an otologist for follow-up care due to the change in hearing and abnormal reflex decay at 1000 Hz for the right ear since the previous audiometric evaluation. The otologist ordered Auditory Braintsem Response (ABR) testing to rule out central findings.

29.3 Auditory Brainstem Response

The ABR was completed using rarefaction polarity click stimuli at 11.1 and 67.1 clicks per second at 90 dB nHL. Results revealed (1) normal absolute latencies for Wave III bilaterally, (2) delayed absolute latencies for Wave V bilaterally, (3) normal interpeak latencies for Waves III through V bilaterally, (4) absent waveforms for Wave I and Wave II for the right ear, (5) absent Wave I for the left ear, (6) normal shift in wave V with increased stimulus repetition rate bilaterally, and (7) normal wave V interaural latency differences. Finally, Wave V on the left ear was delayed slightly more than would be expected based on the degree of hearing loss at 4000 Hz. Although central dysfunction is not strongly indicated in the left ear, it could not be ruled out. Central dysfunction was not suggested for the right ear. The patient was seen by his otologist to discuss the results, also reviewing computed tomographic (CT) scan that revealed no metastasis to bone or evidence of brain lesions. It was recommended that the patient return in 6 months for a repeat audiological examination and follow-up care.

29.3.1 When Are Differences in WRS from One Audiometric Examination to Another Sufficient to Warrant a Referral?

The primary focus of this case report is to help the reader make better decisions regarding what constitutes a significant change in WRS that justifies a referral to another healthcare professional or perhaps leads to a conclusion that a treatment improved or did not improve hearing. For example, Stachler et al reported that many otolaryngologists regard an improvement of 10 to 20% in WRS as evidence that corticosteroid treatments such as prednisone, methylprednisolone, solumedrol, and dexamethasone delivered via the oral, intravenous, and/or intratympanic (IT or "salvage treatment") routes for inner ear disorders were effective in improving hearing. To the authors of this case report, using a fixed 10 to 20% criterion as evidence of the effectiveness of treatment or change in hearing is worrisome. Based on interactions with students and clinicians over the years, however, it appears that this fixed 10 to 20% criterion is also used by some audiologists as a criterion to determine significant change in hearing.

The authors' concern is that the use of an arbitrary fixed criterion is not appropriate based on the research of Thornton and Raffin and Carney and Schlauch. Results of these investigators are available to help clinicians determine if significant differences between WRSs are present or whether differences are simply due to the variability of the test measures. Thornton and Raffin created a theoretical framework based on the assumption that a response to a stimulus is independent of a response to another stimulus. The authors viewed word recognition test results as being binomially distributed and used statistics to describe their characteristics. That is, WRSs have greater variability in the middle range and less variability in the upper and lower ranges. In addition, variability across the entire range decreases as the number of words that are administered increases. Carney and Schlauch updated the Thornton and Raffin data using more powerful and sophisticated computers. Table 29.2 reports the lower and upper limits of the 95%

Table 29.2 Lower and upper limits of the 95% critical differences for Word Recognition Scores (in %) for a 50-word list.

% Score	Thornton and Raffin (1978)	Carney and Schlauch (2007)
0	0–4	0–6
2	0–10	0–10
4	0–14	0–14
6	2–18	0–18
8	2–22	2–20
10	2–24	2–24
12	4–26	4–26
14	4–30	4–28
16	6–32	6–32
18	6–34	6–34
20	8–36	8–36
22	8–40	10–38
24	10–42	10–42
26	12–44	12–44
28	14–46	14–46
30	14–48	16–48
32	16–50	16–50
34	18–52	18–52
36	20–54	20–54
38	22–56	22–56
40	22–58	24–58
42	24–60	24–60
44	26–62	26–62
46	28–64	28–64
48	30–66	30–66
50	32–68	32–68
52	34–70	34–70
54	36–72	36–72
56	38–74	38–74
58	40–76	40–76

Table 29.2 continued

% Score	Thornton and Raffin (1978)	Carney and Schlauch (2007)
60	42–78	42–76
62	44–78	44–78
64	46–80	46–80
66	48–82	48–82
68	50–84	50–84
70	52–86	52–84
72	54–86	54–86
74	56–88	56–88
76	58–90	58–90
78	60–92	62–90
80	64–92	64–92
82	66–94	66–94
84	68–94	68–94
86	70–96	72–96
88	74–96	74–96
90	76–98	76–98
92	78–98	80–98
94	82–98	82–100
96	86–100	86–100
98	90–100	90–100
100	96–100	94–100

Note: Values within each range are not significantly different from the value reported to the left under ("%score"). The second column lists the data from Thornton and Raffin (1978), and the third column lists the data from Carney and Schlauch (2007). Note: Values in bold, italics, and underlined are those that are different from the values originally reported by Thornton and Raffin (1978).

critical differences for WRSs (in %) for a 50-word list. Values within each range are not significantly different from the value reported to the left under "%score." The second column lists the data from Thornton and Raffin and the third column lists the data from Carney and Schlauch. Values in bold, italics, and underlined are those that were found by Carney and Schlauch to be different from the values originally reported by Thornton and Raffin. Carney and Schlauch found that 14 of the 51 possible ranges were different (27.5%) with the range expanded in 4 instances and contracted in 10 instances.

When determining if differences in WRSs are significant, clinicians should consult Table 29.2 to compare changes in WRSs from one audiological examination to another (e.g., annual audiograms, ototoxic monitoring) or from pre- to post-treatment. As may be seen in Table 29.1, the patient's WRS on 3/11 for the right ear using a recorded female talker 50-word list was 80% and it decreased to 40% on 3/12 (i.e., a decrease of 40%). For the left ear, the WRS on 3/11 was 82% and on 3/12 it was 70% (i.e., a decrease of 12%). In looking at Table 29.2, the initial score of 80%, the follow-up WRS must be poorer than 64% for the difference to be significant. In this case, the follow-up WRS was 40%; thus this 40% decrease in WRS is significant ($p < 0.05$) and not related to chance. On the other hand, for the left ear, in looking at Table 29.2, with the initial score of 82% the follow-up WRS must be poorer than 66% for the difference to be significant. In this case, the follow-up WRS was 70%; thus this 12% decrease in WRS is not significant ($p > 0.05$). When using Table 29.2, whether using the data from Thornton and Raffin or Carney and Schlauch, it is clear that the decrease in the right ear is significant, whereas the decrease for the left ear is not. If the clinician had used the criterion of a fixed 10% for the left ear, then he or she might have referred this patient for follow-up care, which could have led to unnecessary anxiety, expense, and testing.

Another, more atypical, comparison is to compare changes in WRS over time. In this patient, the best WRS was 92% for the right ear and 98% for the left ear, whereas the poorest WRS was 40% in the right ear and 70% for the left ear. In viewing Table 29.2, one may see that differences indicate a significant ($p < 0.05$) decrease bilaterally. In looking at Table 29.1 the WRS for the right ear gradually declined from 92% in 3/09 to 80% in 3/11. For the left ear, the WRS was unchanged from 3/07 to 3/09 (98% to 92%). In 3/10 the WRS for the left ear decreased to 88%, which would have met the criterion of $p < 0.05$ if the comparison was to the 98% in 3/07 and 3/08. The change would not be significant if the comparison was to 3/09 (i.e., 92%). Therefore, the left ear comparisons from year to year would never have met the criteria of significant differences. If the comparisons were made between 3/12 (70%) and any other year than 3/11 (82%), the decrease in WRS would be interpreted as significant.

29.4 Questions to the Reader

1. What is judged a significant decrease in hearing to warrant referral to an otologist?
2. Is it better to recommend referral to a general otolaryngologist or to an otologist in these cases?
3. The patient did not report tinnitus in either ear, and the results of tympanometry, acoustic reflex thresholds, and acoustic reflex decay were consistent with cochlear involvement. Do you think a referral to an otologist was necessary?

29.5 Discussion of Questions to the Reader

1. What is judged a significant decrease in hearing to warrant referral to an otologist?

   At our clinic, a decrease in hearing level of 10 dB or greater at more than one frequency in either ear from the previous audiogram is considered significant. In addition, a decrease in a WRS is considered significant if the WRS at the follow-up visit exceeds the previous WRS using the binomial distribution (Carney and Schlauch column in Table 29.2). To determine this, the audiologist would look at Table 29.2 from Carney and Schlauch to compare the follow-up WRS to the previous WRS. Table 29.2 from Carney and Schlauch is placed at each audiometer in the authors' clinic for easy retrieval and interpretation. For example, in this case, the patient's previous WRS in the right ear was 80%, and the WRS for the follow-up visit was 40% Using the 50-word list column in Table 29.2, the follow-up WRS must be poorer than 64% to be considered a significant decrease ($p < 0.05$). In this case, the follow-up WRS was 40% and thus the decrease in WRS was judged significant. For the left ear, a change from 82 to 70% was found not to be significant.

2. Is it better to recommend referral to a general otolaryngologist or to an otologist in these cases?
   The authors believe a referral to an otologist is more appropriate than a referral to a general otolaryngologist if there is concern about the integrity of any segment of the auditory system.
3. The patient did not report tinnitus in either ear, and the results of tympanometry, acoustic reflex thresholds, and acoustic reflex decay were consistent with cochlear involvement. Do you think a referral to an otologist was necessary?
   Yes, a referral to an otologist was necessary due to the significant decrease in hearing levels and WRS. In addition to making a recommendation to the patient's primary care physician for a referral to an otologist, the audiologists also recommended that ABR testing be completed.

29.6 Outcome

Due to the 10 dB decrease in hearing in the right ear at 3000 to 4000 Hz, the patient's right hearing aid was reprogrammed to accommodate the change. At the conclusion of the fit, the patient stated that his word recognition had improved slightly. Also, he continues to see his otolaryngologist to determine if any further care may be implemented to improve the swelling of his right ear canal. Finally, the patient is scheduled to be seen in 6 months by his otologist and audiologist to monitor any changes in thresholds or WRS. The patient has been counseled on the availability of Hearing Assistive Technology to supplement the performance provided by his hearing aids, but for now that recommendation has not been followed. Finally, due to the poor WRS in the right ear and possible decrease in the future, some conversation has begun with the patient concerning the possible benefits of a cochlear implant. For now he has expressed a lack of interest in any type of surgical intervention.

29.7 Key Points

1. Develop a Best Practice Guideline for your clinic to include the following:
   a) Use of full lists and recorded speech material when evaluating WRS. Do not use half lists or monitored live voice.
   b) Protocols for when follow-up audiological examinations indicate significant changes in hearing and what recommendations should be made.
2. Use the evidence to make sound clinical judgments and not some arbitrary criterion that seems to be passed from one person to another without solid backing from the evidence cited in the literature.
3. The reader should retrieve, read, and refer back to the articles by Thornton and Raffin and Carney and Schlauch.

Suggested Reading

Carney E, Schlauch RS. Critical difference table for word recognition testing derived using computer simulation. J Speech Lang Hear Res 2007; 50: 1203–1209

Stachler RJ, Chandrasekhar SS, Archer SM et al. American Academy of Otolaryngology-Head and Neck Surgery. Clinical practice guideline: sudden hearing loss. Otolaryngol Head Neck Surg 2012; 146 Suppl: S1–S35

Thornton AR, Raffin MJ. Speech-discrimination scores modeled as a binomial variable. J Speech Hear Res 1978; 21: 507–518

30 Revisiting Acoustic Reflexes

Wayne J. Wilson and Alicja N. Malicka

MP is a 38-year old female referred by her ear, nose, and throat (ENT) physician for an audiological examination following sudden onset of bilateral hearing loss.

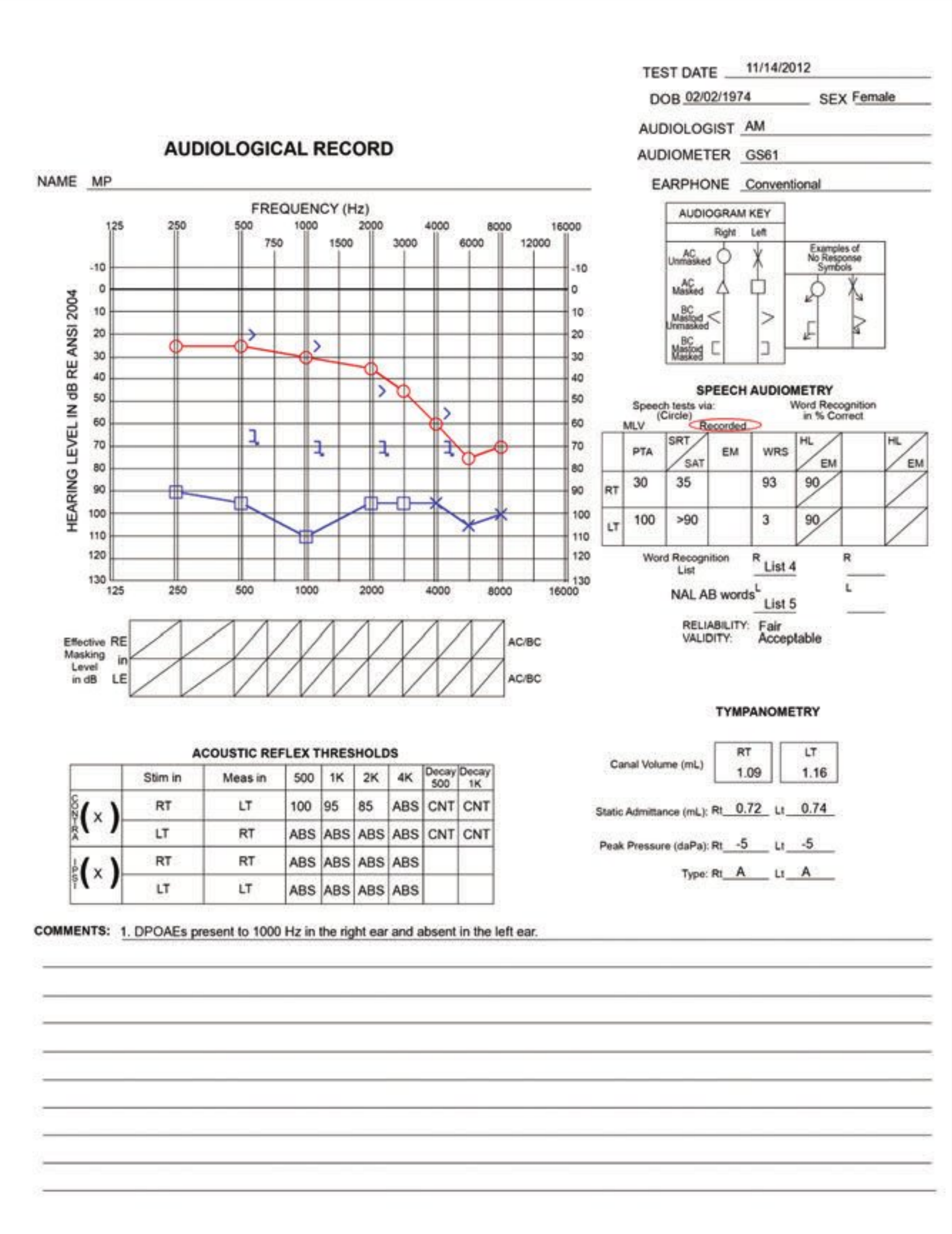
30.1 Clinical History and Description

MP is a 38-year-old female who awoke one morning with bilateral hearing loss. On seeing her family physician that same day, MP was diagnosed with bronchiectasis (abnormal widening of the bronchi or their branches, causing risk of infection) and was immediately referred to an ear, nose, and throat (ENT) physician. MP's ENT physician diagnosed MP as experiencing a bilateral sudden sensorineural hearing loss (SSNHL) caused by ischemic episodes in the cochlear/vestibular arteries. MP's ENT physician started MP on corticosteroids and referred MP to an audiologist for an audiological examination and follow-up monitoring.

MP's audiological case history revealed that for the past week she had been experiencing headaches, pharyngitis, and mild otalgia, which she had been self-managing with bed rest and over-the-counter medications. In the days immediately preceding her bilateral SSNHL, MP experienced occasional high-pitched tinnitus in her left ear and brief (<30 seconds) episodes of vertigo. MP also reported that her father was born with a bilateral mild to moderate sensorineural hearing loss (SNHL) for which he has been successfully fit with hearing aids. The remainder of MP's audiological case history was unremarkable.

30.2 Audiological Testing

Fig. 30.1 reports the results of MP's initial audiological examination performed by her audiologist. Pure-tone testing revealed a mild to severe SNHL that is gradually sloping in configuration in the right ear and a profound SNHL in the left ear. MP's speech recognition threshold (SRT) in the right ear revealed a mild loss


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols

AUDIOLOGICAL RECORD

TEST DATE 11/14/2012
DOB 02/02/1974 SEX Female
AUDIOLOGIST AM
AUDIOMETER GS61
EARPHONE Conventional
NAME MP

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	30	35		93	90		
LT	100	>90		3	90		

Word Recognition List NAL AB words R List 4 L List 5 R L

RELIABILITY: Fair
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.09	1.16

Static Admittance (mL): Rt 0.72 Lt 0.74
Peak Pressure (daPa): Rt -5 Lt -5
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	100	95	85	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS: 1. DPOAEs present to 1000 Hz in the right ear and absent in the left ear.

Fig. 30.1 MP's audiological examination.

in the ability to receive speech and is in agreement with the pure-tone average. The SRT for the left ear revealed a profound loss in the ability to receive speech and is also in agreement with the pure-tone average. Because this word recognition testing was completed in Australia, PB’s WRSs were measured using the National Acoustics Laboratories’ Arthur Boothroyd (NAL-AB) word lists and its recommended protocols. The NAL-AB word lists contain 15 lists of 10 monosyllabic words. The patient’s response to each word is scored phonemically (30 scoreable phonemes per list) such that correctly repeating three, two, one, or none of the phonemes in each word elicits a score of 10%, 7%, 3%, or 0% for that word, respectively. A performance-intensity (PI) function is measured for each ear by presenting the first word list at the expected half-maximum level (HML: the level at which the patient is predicted to score 50%) and subsequent lists at levels 15 dB higher than the previous word list until a maximum word recognition score is reached. If not already found for the first word list, a score is also obtained at a signal level below HML. Speech noise is applied to the nontest ear according to the formula: masking level in nontest ear = presentation level in test ear - 40 dB + the air–bone gap in nontest ear + audiometer conversion factor. Once the PI function is plotted, the SRT is estimated by extrapolating the presentation level required to reach the 50% score on the PI function. Using this protocol, MP’s word recognition scores (WRSs) revealed normal ability to recognize speech in the right ear and very poor ability to recognize speech in the left ear at a presentation level of 90 dB HL. For MP’s right ear, the WRS improved with increased presentation levels to a maximum of 93% with no significant rollover at the 90 dB HL presentation level.

MP’s tympanometry testing showed ear canal volume (mL), static admittance (mL), and middle ear pressure (daPa) values within normal ranges bilaterally. MP’s acoustic reflex thresholds (ARTs) showed the right contralateral ARTs (stimulus tone in the right ear and recording probe in the left ear) were present at 500 to 2000 Hz but the left contralateral ARTs (stimulus tone in the left ear and recording probe in the right ear) and right and left ipsilateral ARTs (stimulus tone and recording probe in the same ear) were absent at all tested frequencies. MP’s distortion-product otoacoustic emissions (DPOAEs) were also shown to be present up to 1000 Hz in the right ear and absent in the left ear.

30.3 Question to the Reader

1. Are MP’s ARTs consistent with her history and audiological test results?

30.4 Discussion of Question to the Reader

1. Are MP’s ARTs consistent with her history and other audiological test results?

   On first inspection, MP’s ARTs appear to be inconsistent with her history and other audiological test results. To identify this inconsistency, we must consider the acoustic reflex pathway and how the potential site(s) of lesion suggested by MP’s history and other audiological test results map onto this pathway.

   Fig. 30.2 reports the ipsilateral and contralateral acoustic reflex pathways as described by Gelfand. The ipsilateral acoustic reflex pathway (stimulus tone and recording probe in the same ear) traverses the cochlea, the eighth cranial nerve (CN VIII: the vestibulocochlear nerve), ventral cochlear nucleus (VCN), superior olivary complex (SOC), facial nerve nucleus (FNN), the seventh cranial nerve (CN VII: the facial nerve), and stapedius muscle on the same side of the head. The contralateral acoustic reflex pathway (stimulus tone and recording probe in opposite ears) traverses the cochlea, CN VIII, VCN, and SOC on one side of the head before crossing over to the SOC, FNN, CN VII, and stapedius muscle on the opposite side of the head. Lesions in any of these anatomical structures could affect an ART whose pathway crosses the affected structure(s).

   MP’s history and other audiological test results sum to suggest the presence of a cochlear (sensory) and/or eighth CN (neural) site of lesion bilaterally. This is because of MP’s reported SSNHL and her audiological results showing a mild to severe SNHL in the right ear and a profound SNHL hearing loss in the left ear, normal tympanograms bilaterally, and

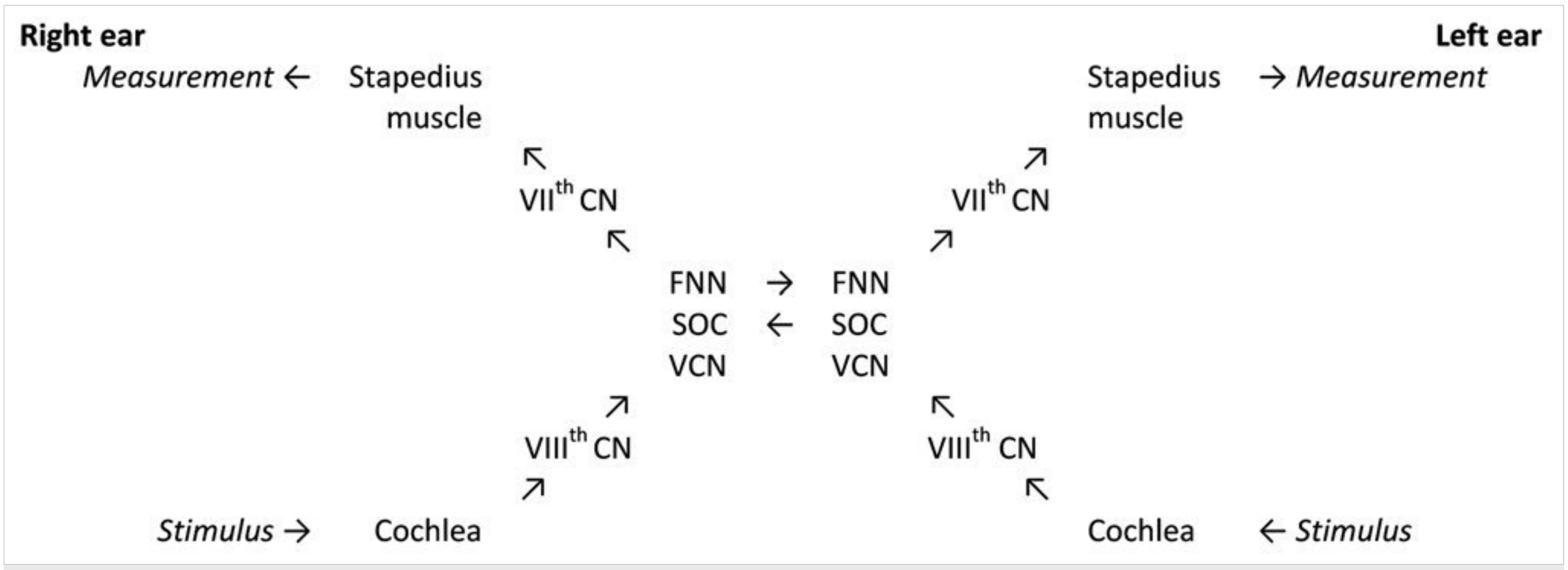


Fig. 30.2 The acoustic reflex threshold pathways according to Gelfand (2009).

present DPOAEs to 1000 Hz in the right ear and absent DPOAEs in the left ear.

Mapping MP's potential bilateral cochlear (sensory) and/or eighth CN (neural) sites of lesion onto Fig. 30.2 does not predict MP's measured ART pattern. That is, these sites of lesion would not have predicted that MP's ARTs would be present only for the right contralateral condition (stimulus tone in the right ear and recording probe in the left ear) at 500 to 2000 Hz. Instead, mapping these sites of lesion onto Fig. 30.2 predicts two other ART patterns. The first predicted ART pattern would be for the ARTs to be present in the right ipsilateral and right contralateral conditions only. This would have been the result if the site of lesion in the right ear was the cochlea (with the mild degree of cochlear hearing loss allowing the right ear's eighth CN to be stimulated) and the site of lesion in the left ear was the cochlea and/or the eighth CN (with the profound cochlear loss preventing the left ear's eighth CN from being stimulated and/or the left ear's eighth CN lesion preventing this nerve from responding). The second predicted ART pattern would be for the ARTs to be absent in all four ART conditions. This would have been the result if the site of lesion in the right ear was the eighth CN (preventing this nerve from responding) and the site of lesion in the left ear was the cochlea and/or the eighth CN (with the profound cochlear loss preventing the left ear's eighth CN from being stimulated and/or the left ear's eighth CN lesion preventing this nerve from responding).

Mapping MP's measured ART pattern onto Fig. 30.2 suggests MP could have a second site of lesion in her right ear that was not identified from the case history or other audiological test results. To reveal an ART pattern where ARTs are only present in the right contralateral condition (stimulus tone in the right ear and recording probe in the left ear), the two sites of lesion in MP's right ear would have to be the cochlea and the seventh CN (with the mild degree of cochlear hearing loss allowing the right ear's eighth CN to be stimulated and the right ear's seventh CN lesion preventing this seventh CN from responding). The site of lesion in MP's left ear would still have to be the cochlea and/or the eighth CN (with the profound cochlear loss preventing the left ear's eighth CN from being stimulated and/or the left ear's eighth CN lesion preventing this nerve from responding). This combination of lesions would leave the right contralateral acoustic reflex pathway (stimulus tone in the right ear and recording probe in the left ear) as the only pathway not to traverse a lesion capable of eliminating ARTs.

It is worth noting that replacing the right seventh CN site of lesion suggested earlier with a right mild conductive site of lesion would have resulted in the same ART pattern revealed for MP. Although such a mild conductive site of lesion was possible given MPs recent headaches, pharyngitis, and mild otalgia, it is improbable given that MP presented with no history of an outer or middle ear disorder, and her right ear revealed no air–bone gaps. In addition, DPOAEs were present to 1000 Hz, and a type A tympanogram was recorded.

30.5 Diagnosis and Recommended Treatment

Overall, MP's case history and audiological examination results were most consistent with cochlear and seventh CN sites of lesion on the right side and a cochlear site of lesion on the left side; however, an eighth CN site of lesion on the left side cannot be ruled out.

With MP's SSNHL already being aggressively managed by her ENT physician, the audiologist decided to further investigate the possible right-side seventh CN by once again asking MP if she had experienced any event in the past that could have affected her head, neck, and/or jaw. When questioned once again, MP revealed that 10 years prior to her SSNHL she had successfully undergone surgery on her right temporomandibular joint (TMJ: the joint connecting the mandible to the skull) to alleviate chronic pain she had been experiencing in and around that joint. When asked if there had been any complications, MP stated she had experienced right-sided facial palsy after this surgery, although she believed she had quickly and fully recovered from this palsy. This new information, plus previous reports in the literature of seventh CN complications following surgical procedures for the treatment of TMJ, supported the right-side seventh CN site of lesion suggested by MP's audiological test results.

In addition to reporting the audiological results and scheduling to monitor MP's hearing on a weekly basis while MP was under the care of her ENT physician, the audiologist also reported the possible right-side seventh CN site of lesion to MP's ENT physician. The audiologist offered to assess MP's seventh CN function via electroneurography (EnoG). In addition to accepting the audiologist's suggestion, MP's ENT physician included the possible involvement of MP's seventh CN in the physician's management of MP's SSNHL.

30.6 Key Points

1. By completing ipsilateral and contralateral ART measures, the audiologist was able to assess all components of the acoustic reflex pathways.
2. By mapping the audiological results on to the acoustic reflex pathways shown in Fig. 30.2 and by rigorous history taking, the audiologist was able to identify a potential seventh CN site of lesion that would otherwise have remained undetected.

Suggested Reading

Gelfand SA. The acoustic reflex. In: Katz J, Medwetsky L, Burkard R, Hood L, eds. Handbook of Clinical Audiology. 6th ed. Baltimore, MD: Lippincott Williams & Wilkins; 2009:189–221

do Egito Vasconcelos BC, Bessa-Nogueira RV, da Silva LC. Prospective study of facial nerve function after surgical procedures for the treatment of temporomandibular pathology. J Oral Maxillofac Surg 2007; 65: 972–978

31 Could It Be (Central) Auditory Processing Disorder?

Wayne J. Wilson

JS is a 25-year-old male receiving service from a brain injury rehabilitation unit (BIRU) who was referred by his social worker for a (central) auditory processing CAP assessment in preparation for his potential return to work.

31.1 Clinical History and Description

JS is a 25-year-old male who has spent the last 6 months in the BIRU of a large, adult hospital following a traumatic brain injury (TBI) he incurred during a motor vehicle accident.

At the scene of the accident, JS was found to be disoriented, walking with an unsteady gait, and hemorrhaging (bleeding) from both ears. On arrival at the hospital, JS underwent many assessments, including an assessment of his consciousness using the Glasgow Coma Scale (GCS). The GCS is a neurological scale with three elements. The first examines eye response, with scores ranging from 1 (does not open eyes) to 4 (opens eyes spontaneously). The second examines verbal response, with scores ranging from 1 (makes no sounds) to 5 (oriented, converses normally). The third examines motor response, with scores ranging from 1 (makes no movements) to 6 (obeys commands). These three component scores are summed to calculate a final GCS score ranging from 3 (indicating deep unconsciousness) to 15 (indicating normal consciousness). On arrival at the hospital, JS's GCS score was 15 (eye opening = 4, opens eyes spontaneously; verbal = 5, orientated, converses normally, motor = 6, obeys commands), but within 20 minutes this had declined to 6 (eye opening = 1, does not open eyes; verbal = 2, incomprehensible sounds, motor = 3, abnormal flexion to painful stimuli). He also started to experience bradycardia (slow heart rate), anisocoria (unequal pupils), and intermittent seizure activity. Fearing JS had sustained a TBI, JS's emergency room physicians requested an urgent computed tomographic (CT) head scan, which revealed bifrontal and bitemporal contusions (bruises) with associated subarachnoid hemorrhage (SAH—bleeding on the brain) and a left-side subdural hematoma (SDH—a collection of blood on the brain) with a right midline shift (the brain had been pushed to the right). A bifrontal craniectomy (removal of parts of the skull) was performed to evacuate the SDH and alleviate the intracranial pressure, during which JS's subfrontal brain regions were found to be swollen and severely contused. JS was then transferred to a neurosurgical intensive care unit (NICU) for further treatment and monitoring using a wide range of drugs (including diuretics, antiseizure drugs, coma-inducing drugs, and anticonstipation drugs) and physiological measures (including pulse, heart rate, breathing rate, arterial blood oxygen and carbon dioxide saturations, and intracranial pressure). Over the course of the next 2 weeks, JS showed frequent hypotensive (low blood pressure) events with resulting decreases in cerebral perfusion pressure (low blood pressure in the brain) leading to an infarct (obstructed blood supply) in the left temporoparietal region of his brain. Fortunately, JS also showed a reversal of his midline shift, a slow improvement in his GCS score to 10 (eye opening = 2, opens eyes in response to painful stimuli; verbal = 2, incomprehensible sounds, motor = 6, obeys commands), and he progressed to a mechanical soft diet.

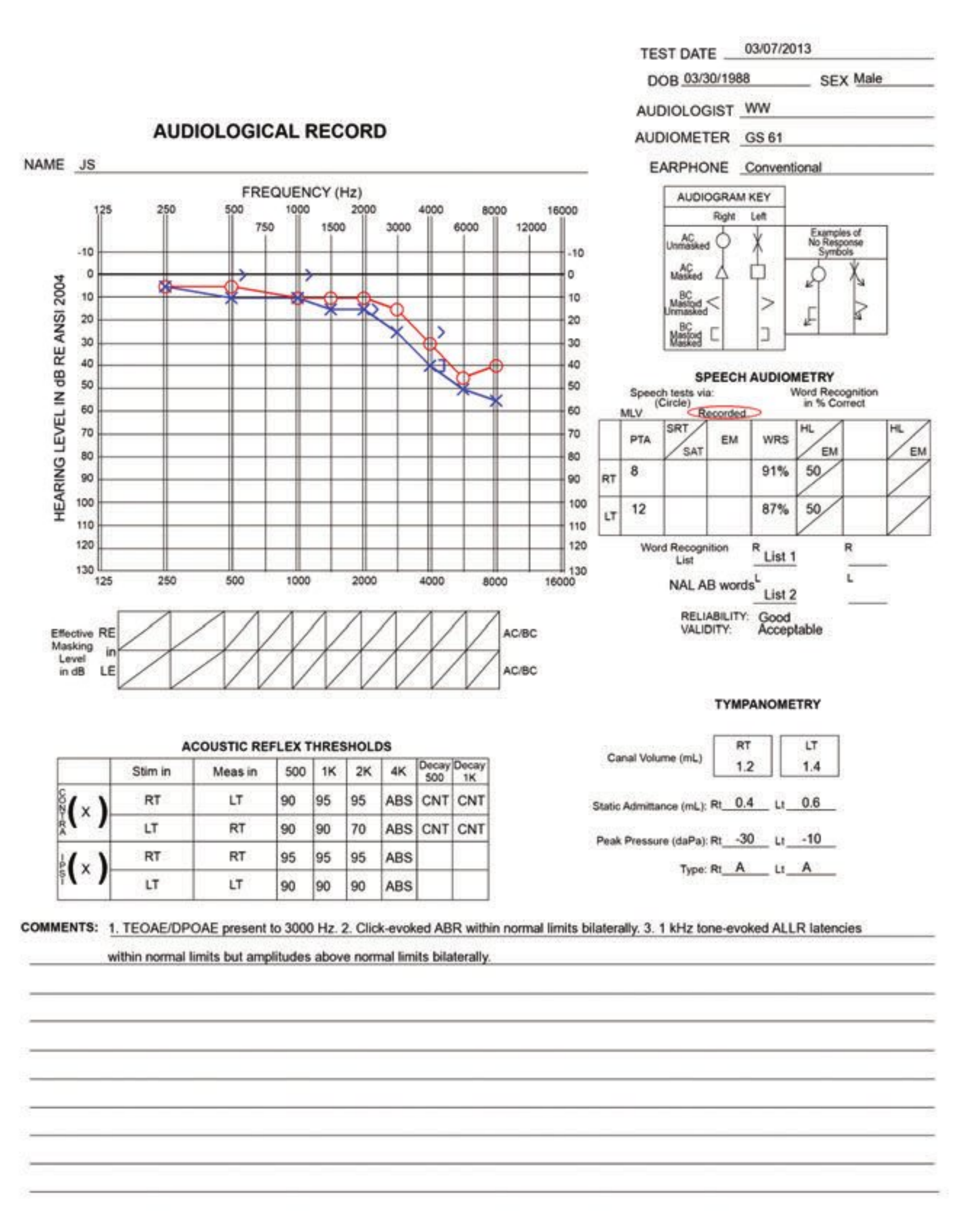
JS was transferred as an inpatient to the BIRU in the third week after his TBI. This BIRU contained its own inpatient ward, an independent living unit, and a "day hospital," all aimed at providing a multidisciplinary, progressive management pathway for patients with TBI. Over the subsequent 6 months, JS's GCS score returned to 15 and he was able to undergo a range of assessments and rehabilitation programs under the care of his physicians, physical therapists, neuropsychologists, speech-language pathologists, social workers, and audiologists.

JS's team at the BIRU now consider him ready for discharge, and his social worker is preparing the way for JS's potential return to work. JS was previously employed as a help desk operator for an information technology (IT) company where he solved client IT problems over the phone and Internet in a noisy call center. The social worker has multiple concerns about JS's ability to return to this work, including his ability to function in such a noisy environment. To address the latter, she referred JS to the audiology clinic at a large university for a CAP assessment. The referral letter includes a summary of JS's most recent assessments at the BIRU, which include the following:

- Social work: JS has excellent support from his family (mother, father, brother, and girlfriend) who are willing to care for JS on discharge from the BIRU. JS also has excellent support from his current employer who is motivated to see JS return to work for the company.
- Medical: JS's GCS score remains at 15. His most recent CT head scan revealed residual bifrontal, bitemporal, and left temporoparietal lesions in his brain.
- Physical therapy: JS continues to show some reduced fine-motor function (particularly moving his feet), but JS can walk without assistance.
- Neuropsychology: JS continues to show mild levels of anxiety and difficulties concentrating, tracking more than one thing at a time, completing complex tasks (often requiring longer times to do so), learning large amounts of new information, organizing, and self-monitoring (sometimes being verbose and taking conversations "off-track"). JS does, however, have a normal memory for past knowledge, is able to remember moderate amounts of new information, has an average attention span, and is able to problem-solve at an average level.
- Speech-language pathology: JS continues to show difficulties with comprehension, abstract meanings, idioms, proverbs, nonspecific references, confabulation, word-finding, topic maintenance, and judging context. JS also has minor difficulties initiating speech and controlling his overall rate of speech.

31.2 Audiological Testing

Fig. 31.1 reports the results of JS's most recent audiological examination, which was completed 1 week prior to his behavioral [C]AP assessment that is yet to be described here. JS


FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
Effective Masking Level in dB
RE
in
LE
AC/BC
AC/BC

AUDIOLOGICAL RECORD

NAME JS

TEST DATE 03/07/2013
DOB 03/30/1988 SEX Male
AUDIOLOGIST WW
AUDIOMETER GS 61
EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT	8			91%	50		
LT	12			87%	50		

Word Recognition List R List 1 R
NAL AB words L List 2 L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	90	95	95	ABS	CNT	CNT
	LT	RT	90	90	70	ABS	CNT	CNT
IPSI (x)	RT	RT	95	95	95	ABS		
	LT	LT	90	90	90	ABS		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.4

Static Admittance (mL): Rt 0.4 Lt 0.6
Peak Pressure (daPa): Rt -30 Lt -10
Type: Rt A Lt A

COMMENTS: 1. TEOAE/DPOAE present to 3000 Hz. 2. Click-evoked ABR within normal limits bilaterally. 3. 1 kHz tone-evoked ALLR latencies within normal limits but amplitudes above normal limits bilaterally.

Fig. 31.1 JS's audiological examination conducted 1 week prior to his behavioral CAP assessment.

presented with a bilateral asymmetrical mild to moderate sensorineural hearing loss (SNHL) at 3000 to 8000 Hz with hearing slightly poorer in the left ear. JS's word recognition scores (WRSs) were consistent with his pure-tone thresholds. Because this testing was completed in Australia, JS's WRSs were obtained using the National Acoustics Laboratories' Arthur Boothroyd (NAL-AB) word lists and its recommended protocols. The NAL-AB word lists contain 15 lists of 10 monosyllabic words. The patient's response to each word is scored phonemically (i.e., 30 scoreable phonemes per list) such that correctly repeating three, two, one, or none of the phonemes in each word elicits a score of 10%, 7%, 3%, or 0% for that word, respectively. Only one NAL-AB word list was presented to each of JS's ears. Speech noise was applied to the nontest ear according to the formula: masking level in nontest ear = presentation level in test ear - 40 dB + air-bone gap in nontest ear + audiometer conversion factor. JS's tympanometry results and contralateral and ipsilateral acoustic reflex thresholds (ARTs) were within normal limits with the exception of absent ARTs at 4000 Hz to contralateral and ipsilateral stimulation, and his transient and distortion-product otoacoustic emissions (TEOAEs and DPOAEs) were present to 3000 Hz. CJ's click-evoked auditory brainstem responses (ABRs) were within normal limits, and his 1000 Hz tone-burst evoked auditory late latency responses (ALLRs) revealed normal latency, but abnormally large amplitude, which were reported as being "suggestive of cortical disinhibition."

31.3 Questions to the Reader

1. Is it possible to complete a behavioral CAP assessment on JS, particularly in light of JS's peripheral hearing loss and his cognitive and language status?
2. How might an audiologist modify the behavioral CAP assessment to better inform the social worker's efforts to return JS to work?

31.4 Discussion of Questions to the Reader

1. Is it possible to complete a behavioral CAP assessment on JS, particularly in light of JS's peripheral hearing loss and his cognitive and language status?

At first glance, the answer would appear to be "no." CAPD has been defined as "a deficit in neural processing of auditory stimuli that is not due to higher order language, cognitive, or related factors," although this definition has recently been challenged. Although JS's most recent CT head scan and auditory evoked potential results suggest CAPD is possible (or even likely), it would be difficult to control for JS's asymmetrical SNHL and his many higher-order cognitive, language, and related factors in a manner that would allow the results of JS's behavioral CAP assessment to be attributed directly to his "true" CAP abilities.

At second glance, a CAP assessment would be possible if the purpose of the assessment is more closely considered. JS's neurological lesions are well documented, so the need to identify the site of lesion is no longer a priority. In fact, JS has been referred by his social worker not for a site of lesion assessment, nor necessarily to confirm or deny the presence of a CAPD, but rather as part of her efforts to return JS to work and to address her concerns about JS's ability to function in a noisy work environment. As a result, the need to identify JS's functional status takes priority, with the audiologist being well placed to determine how JS might perform under different listening conditions.

2. How might you modify your behavioral CAP assessment to better inform the social worker's efforts to return JS to work?

   A typical behavioral CAP assessment involves the patient attending to a series of behavioral CAP tests with his or her results being compared against those of age-matched, normally functioning persons with normal hearing sensitivity (although some CAP tests, such as dichotic digits and frequency patterns, show some resistance to mild hearing losses). Although this approach would allow JS's performance to be judged against these norms, efforts to return JS to work might be better informed by comparing JS's performance between variations of each CAP test, which would allow JS to act as his own control.

31.5 Additional Audiological Testing

JS was evaluated using a behavioral CAP test battery based on the recommendations of the American Speech-Language-Hearing Association and Bellis. This battery included a low-pass filtered speech (LPFS) test, a competing sentences (CS) test, two-pair dichotic digits (TPDD) test, and a frequency attern (FP) test. Each test was administered using headphones in a standard and a nonstandard format as follows:

a) Low-Pass Filtered Speech (LPFS): For each ear separately, JS was asked to repeat 50 monosyllabic words from equivalent word lists presented at 50 dB HL to the test ear. Half the words were filtered (standard format) and half the words were not filtered (nonstandard format). The words were from the Auditec of Saint Louis recordings of the Northwestern University Auditory Test No. 6 (NU-6) word lists 1C and 2C and had been recorded using a male talker. The filtered words had been low-pass filtered using a 1000 Hz cutoff frequency.

b) Competing Sentences (CS): For each ear separately, JS was asked to repeat 20 sentences presented at 35 dB HL to the test ear. Half the sentences were presented while equivalent competing sentences were simultaneously presented at 50 dB HL to the nontest ear (standard format—dichotic), and half the sentences were presented without these competing sentences (nonstandard format—monotic). The sentences were from the Auditec of Saint Louis recordings that were six to seven words in length and had been recorded with a male talker.

c) Two-Pair Dichotic Digits (TPDD): For both ears simultaneously, JS was asked to repeat 20 sets of four digits. Each set of four digits was presented sequentially as two digit pairs with one digit in each pair presented at 50 dB HL (dial setting) to the right ear while the other digit in that pair was simultaneously presented at 50 dB HL (dial setting) to the left ear (standard format—dichotic). For each ear separately, JS was then asked to repeat another 20 sets of four digits where each set of four digits was also presented sequentially as two digit pairs, but each digit in each pair was presented at 50 dB HL to the right ear only (10 sets of four digits) or to the left ear only (10 sets of four digits) (nonstandard format—monotic). The digits were from the U.S. Department of Veterans Affairs recordings that included the numbers 1 through 10 (except 7) and had been recorded using a male talker.

d) Frequency Pattern (FP): For each ear separately, JS was asked to describe the frequency patterns contained in 25 sets of three tones presented monotically at 50 dB HL (standard format). Also for each ear separately, JS was then asked to hum the frequency patterns contained in a further 25 sets of three tones presented monotically at 50 dB HL (nonstandard format). The frequency patterns were from the U.S. Department of Veterans Affairs recording with each tone in each set of three tones being either 880 Hz or 1,122 Hz.

Fig. 31.2 reports JS's results following the behavioral CAP assessment just described.

31.6 Additional Question to the Reader

1. How do JS's behavioral CAP test results inform the social worker's efforts to return JS to work?

31.7 Discussion of Additional Question to the Reader

1. How do JS's behavioral CAP test results inform the social worker's efforts to return JS to work?

   JS's behavioral CAP test results are in two parts: those obtained using standard test formats (LPFS, CS, TPDD, and FP that had to be described in words) and those obtained using the nonstandard test format (unfiltered speech, sentences without competition, two-pairs of digits presented monotically, and frequency patterns that had to be described by humming). JS's results obtained using the standard test formats reveal scores below normal limits on all tests of CAP (the normal limits being the shaded ranges in Fig. 31.2). It remains difficult to directly attribute these scores to CAP disorder, however, in light of JS's asymmetrical high-frequency

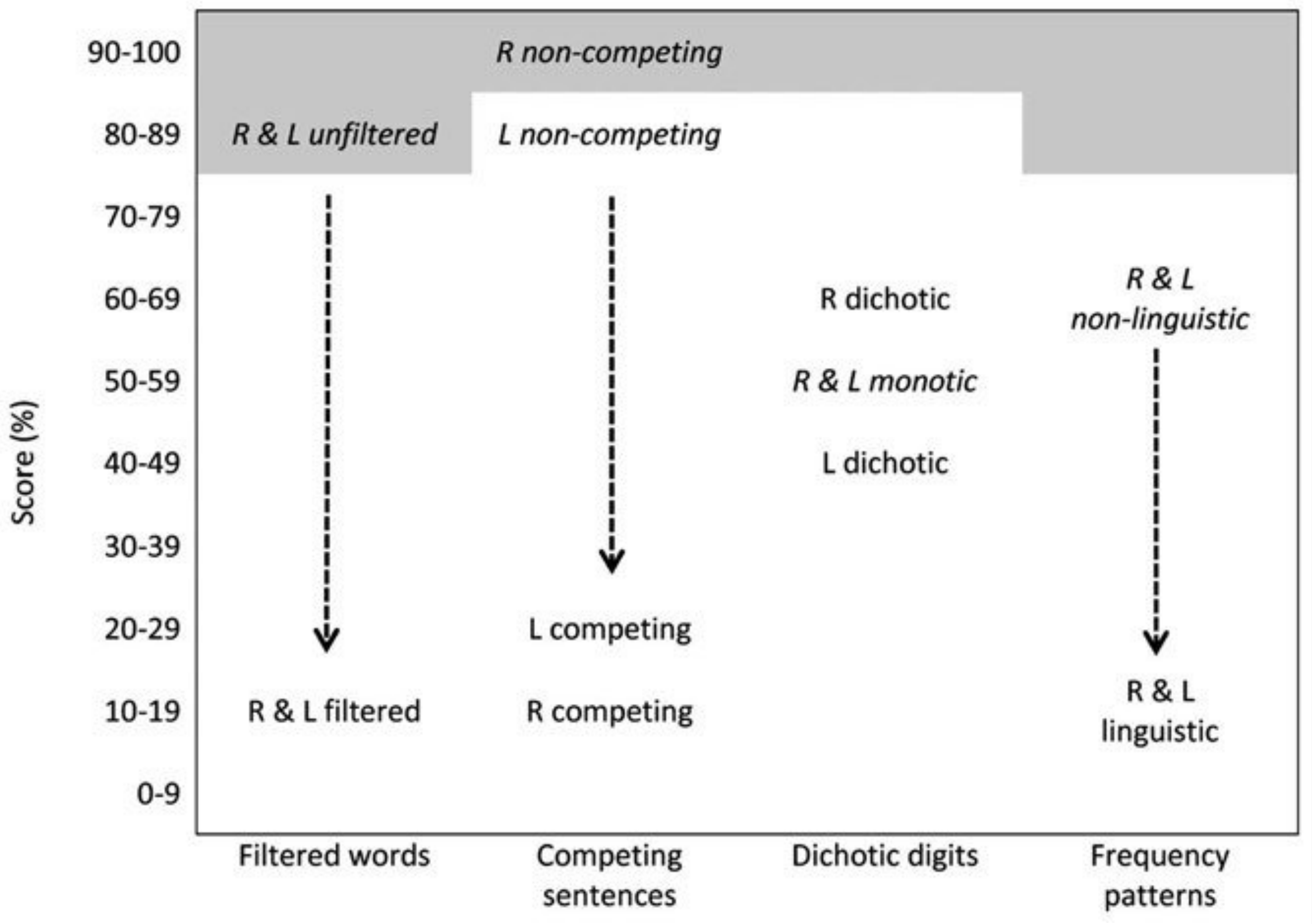


Fig. 31.2 JS's right (R) and left (L) ear scores for each behavioral CAP test applied in the standard (regular text with normal ranges [mean ± 2 SD] shaded) and nonstandard (italicized text with no normal ranges) manner

SNHL and his many higher-order cognitive and language difficulties. By comparing these scores to those obtained using the nonstandard test format, however, it can be seen that JS's ability to repeat monosyllabic words decreased by > 60% when the words were low-pass filtered, JS's ability to repeat sentences decreased by > 50% in the presence of competing sentences, and JS's ability to follow temporal patterns decreased by > 40% when JS had to label those patterns using words. Interestingly, JS's ability to repeat digit pairs did not differ in the dichotic versus monotic conditions. These results suggest JS could face substantial difficulties returning to his previous work as an IT help desk operator in a noisy call center, although caution should always be taken when extrapolating such audiological test results to potential "real world" performance.

31.8 Diagnosis and Recommended Treatment

JS's social worker was able to use JS's CAP test results and his other health-care test results to successfully argue that JS should not return to his previous work as an IT help desk operator. In consultation with JS, his family, his employer, his health-care team, and his newly appointed occupational therapist, JS was instead able to return to work as an IT support officer. This required JS to work in a quiet laboratory solving client IT problems offline in a serial manner. In this setting, JS was able to consult with his colleagues face to face and use assistive listening devices (including a wireless headset that he could connect to his phones, computers, and a roving microphone) in an unhurried manner to improve the signal to noise ratio (SNR) and to slow down the rate of information flow in many of his conversations with coworkers. JS also continued with a range of rehabilitation programs, including CAPD intervention with his audiologist. He continues to recover from his TBI slowly, but steadily.

31.9 Key Points

1. The purpose of conducting a behavioral CAP assessment can vary depending on the individual aspects of the case. The purpose of JS's CAP assessment was functional rather than diagnostic.
2. By modifying the CAP testing, JS's audiologist was able to have JS act as his own control. This at least partly mitigated JS's language and cognitive deficits and allowed the audiologist to better determine JS's functional hearing status.
3. JS's CAP test results contributed to him being reassigned to a job better suited to his current functional status.

Suggested Reading

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Part 3

Diagnostic Examination—Vestibular Function

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32 Diagnosis and Treatment of Superior Semicircular Canal Dehiscence Syndrome

Kelly H. Barninger and Michael J. Ruckenstein

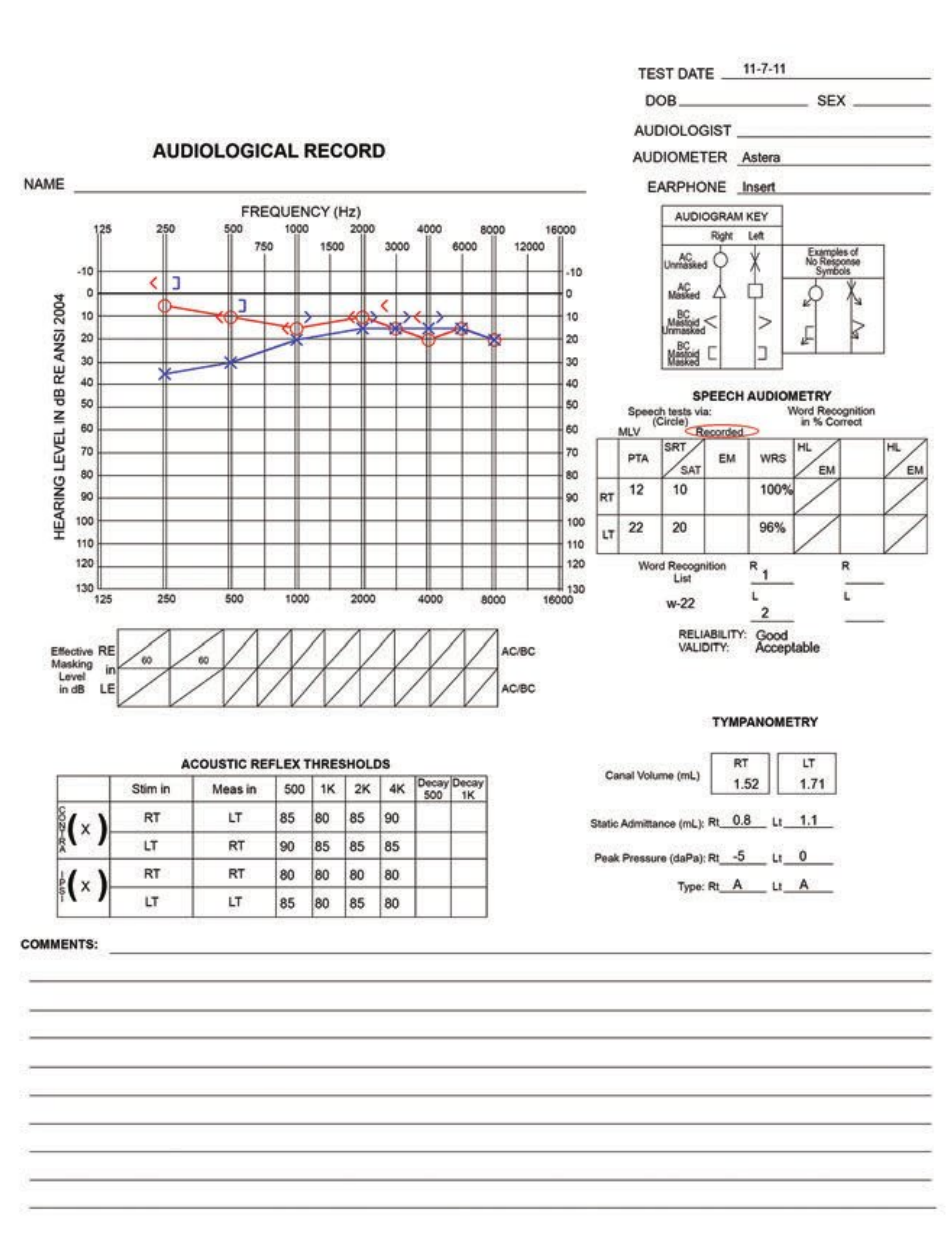
KH is a 49-year-old male who was referred for audiologic and otologic examination. KH was referred based on his report of dizziness provoked by loud sounds as well as aural fullness and tinnitus in the left ear.

32.1 Clinical History and Description

KH presented reporting a several month history of dizziness, imbalance, and lightheadedness lasting seconds to minutes and exacerbated by loud sounds. Also, KH reported brief vertigo associated with positional changes in bed for the past year that resolved in the past few months with Epley maneuvers. Associated symptoms include hearing loss and tinnitus (left greater than right), left autophony, and intermittent aural fullness in the left ear. Symptoms began after an upper respiratory infection and bronchitis. He denied a history of migraines, anxiety, chronic otitis media, previous ear surgeries, or head trauma. KH used a musician's earplug in the left ear to help prevent dizziness and lightheadedness.

32.2 Audiological and Vestibular Testing

An audiologic examination, shown in Fig. 32.1, revealed normal hearing bilaterally, with the exception of a mild conductive hearing loss at 250 and 500 Hz in the left ear. Tympanometry revealed normal middle ear pressure, ear canal volume, and static admittance bilaterally. Acoustic reflex thresholds (ARTs) were present at normal hearing levels for ipsilateral and



AUDIOLOGICAL RECORD

TEST DATE 11-7-11
DOB
SEX
AUDIOLOGIST
AUDIOMETER Astera
EARPHONE Insert
NAME

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	12	10		100%			
LT	22	20		96%			

Word Recognition List w-22: R 1, L 2; R, L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	85	80	85	90		
	LT	RT	90	85	85	85		
IPSI (X)	RT	RT	80	80	80	80		
	LT	LT	85	80	85	80		

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.52	1.71

Static Admittance (mL): Rt 0.8 Lt 1.1
Peak Pressure (daPa): Rt -5 Lt 0
Type: Rt A Lt A

COMMENTS:

Fig. 32.1 Audiological results for KH.

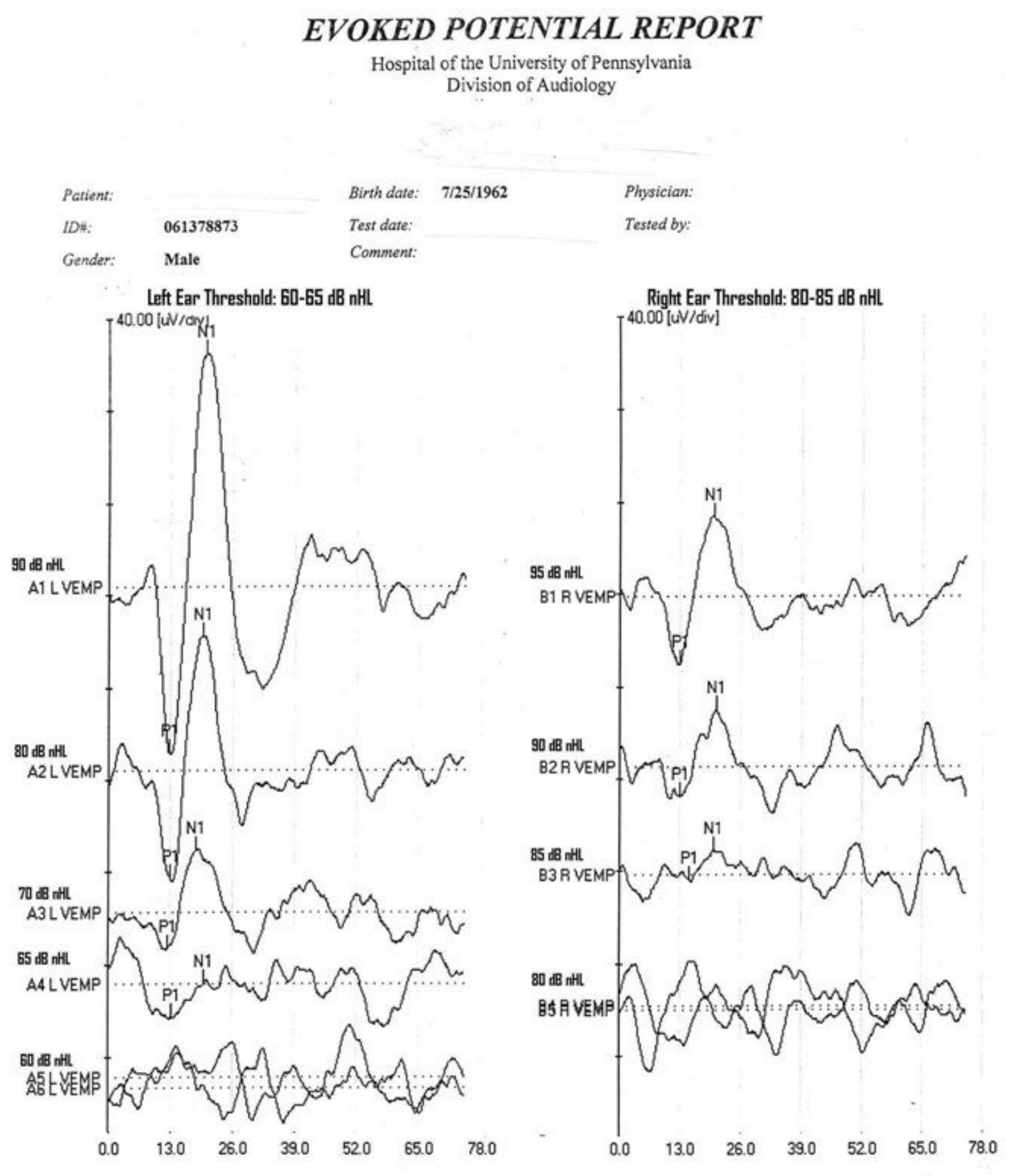
contralateral conditions bilaterally. Word recognition scores (WRSs) were assessed at 50 dB HL and revealed normal ability to recognize speech bilaterally. The speech recognition threshold (SRT) was 10 dB and 20 dB HL for the right and left ear, respectively. The SRTs are in good agreement with the pure-tone average (PTA) bilaterally and indicate normal ability to receive speech in the right ear and a slight loss in the left ear.

As part of an initial examination, three vestibular measures were performed: computerized dynamic posturography (CDP), rotary chair (RC), and videonystagmography (VNG). During CDP testing, the sensory organization test (SOT) and the motor control test (MCT) were completed. The SOT is used to evaluate the patient's ability to effectively use somatosensory, visual, and vestibular cues, either independently or in combination, to maintain postural control. The MCT is used to evaluate the patient's ability to use the automatic motor system to maintain stance and recover from unexpected induced disturbances of increasing size (small, medium, and large platform movements scaled to the height of the patient). The patient's performance was within normal limits for his age in all six conditions of the SOT and in all three conditions of the MCT. Therefore, it was determined that the patient's ability to maintain upright stance under a variety of changing sensory input conditions and to coordinate lower limb and upper body reaction to induced backward sway were normal.

RC testing was completed to evaluate the function of the patient's horizontal semicircular canal vestibulo-ocular reflex (VOR). During the slow-harmonic acceleration test, the patient underwent sinusoidal oscillations at 0.01, 0.04, 0.16, and 0.32 Hz. Results demonstrated normal phase, gain, and asymmetry from 0.01 to 0.32 Hz.

Throughout VNG, performance during oculomotor testing, including smooth pursuit, saccade, and gaze testing, were normal. There was no nystagmus observed throughout spontaneous, post-headshake, hyperventilation, Dix-Hallpike, or position testing. Bithermal caloric responses were considered to be robust and symmetric.

A referral to a neurotologist was scheduled, and, subsequent to examination by the neurotologist, cervical vestibular-evoked myogenic potential (cVEMP) testing and a computed axial tomographic (CT) scan of the temporal bone were ordered to evaluate for superior semicircular canal dehiscence syndrome (SSCD). SSCD was suspected based on the report of dizziness exacerbated by loud sounds, left autophony and aural fullness, and documented low-frequency conductive hearing loss in the presence of normal middle ear function. While cVEMPs were developed to evaluate the function of the saccules and the status of the inferior vestibular nerves, cVEMP has been reported to be particularly helpful in the diagnosis of SSCD. cVEMP testing results, shown in Fig. 32.2, demonstrated an identifiable


EVOKED POTENTIAL REPORT
Hospital of the University of Pennsylvania
Division of Audiology

Patient:
ID#: 061378873
Gender: Male
Birth date: 7/25/1962
Test date:
Comment:
Physician:
Tested by:

Fig. 32.2 Cervical vestibular-evoked myogenic potential results for KH. Note: VEMP response waveforms were repeatable for both ears. For the purposes of figure simplicity, repeated waveforms were not included.

and repeatable P1 and N1 on the summated waveform with thresholds established between 80 and 85 dB nHL for the right ear and between 60 and 65 dB nHL for the left ear. Thresholds were normal in the right ear and abnormal in the left ear.

32.3 Questions to the Reader

1. What is SSCD?
2. What subjective symptoms are commonly reported by patients with SSCD?
3. What are the clinical tests that can be indicative of SSCD?
4. What are the causes of SSCD?

32.4 Discussion of Questions to the Reader

1. What is SSCD?
   SSCD is a defect of the otic capsule of the vestibular labyrinth, specifically involving the most superior aspect of the superior semicircular canal. Due to this bony deficiency, the endosteum of the superior canal interfaces directly with the dura of the temporal lobe creating a "third window" within the inner ear. The presence of the third window allows for endolymph movement within the superior canal when the inner ear is subjected to higher levels of sound or pressure stimuli. The resulting cupular deflection causes symptoms of vertigo, imbalance, and nystagmus patterns that are consistent with a superior semicircular canal–mediated process.
2. What subjective symptoms are commonly reported by patients with SSCD?
   Hearing-related symptoms include hearing loss, aural fullness, hyperacusis, and autophony in the affected ear(s). Additionally, the patient may report hearing internal noises, such as eye movements, joint movements, chewing, and heartbeat (pulsatile tinnitus). Balance-related symptoms include sound- or pressure-induced vertigo, oscillopsia (inability to focus on a target), dizziness, and/or chronic disequilibrium.
3. What are the clinical tests that of SSCD?
   The presence of SSCD on a CT scan is insufficient to diagnose the syndrome. This is due to the fact that CT scan tends to provide false-positive results in identifying SSCD. In addition, SSCD is present on a CT scan in a significant number of patients without symptoms of SSCD. Therefore, the clinical diagnosis of SSCD must be corroborated by one or more objective tests in addition to an abnormal CT scan. These additional objective tests can include:
   a) Air–bone gaps, generally low-frequency with normal bone-conduction thresholds, normal tympanogram, and present ARTs. Bone conduction thresholds may even be present at hearing levels better than 0 dB HL.
   b) A cVEMP thresholds below 70 dB nHL on the side of the dehiscence. This low level is considered to be abnormal.
   c) Ocular VEMPS (oVEMPs), in which auditory click stimuli are presented to one ear and the compound action potential (AP) generated by contraction of the ocular muscles is recorded, may provide a superior screening method when compared with cVEMPs. Characteristic abnormalities on oVEMPs include abnormally large amplitude waveforms in response to the click stimulus. That being said, as of the writing of this case report, oVEMP testing is not commercially available.
4. What are the causes of SSCD?
   The underlying dehiscence of the superior canal likely represents a congenital developmental defect of the temporal bone. In some cases, the otic capsule is dehiscent from birth. In other cases, the capsule is abnormally thin and the dehiscence eventually results from bony erosion due to chronic pressure from the overlying brain. In either situation, the presence of the defect is not sufficient to cause symptoms. Symptoms typically occur in adulthood after "activation" of the defect by some traumatic event. Such traumatic events may include direct blows to the head, acoustic trauma, barotrauma from an airline flight, scuba diving, or a strong sneeze or cough.

32.5 Additional Testing

32.5.1 Imaging

A high-resolution CT scan of the temporal bone was ordered by the neurotologist. The radiologist's impressions were that the CT scan revealed thinning and/or dehiscence of the roof of the left superior semicircular canal. Otherwise, the appearance of the temporal bone was unremarkable.

32.6 Additional Questions to the Reader

1. What are treatment options for patients with SSCD?
2. If cVEMPs were abnormal, why were VNG and RC results normal?
3. Why are cVEMP thresholds lower (abnormal) in patients with SSCD, and why are air–bone gaps with normal bone conduction thresholds typically present on the audiogram?

32.7 Discussion of Additional Questions to the Reader

1. What are treatment options for patients with SSCD?
   Counseling the patient to understand SSCD and the importance of avoiding precipitating stimuli may be sufficient in patients with symptoms of aural fullness and mild hearing loss. In those patients with the full spectrum of SSCD, surgical repair of the defect is indicated. Surgical repairs can be divided into techniques that "defunction" the superior canal by plugging it. Such plugging procedures can be accomplished via a middle fossa craniotomy or via a transmastoid approach. Other more conservative procedures have become popular when the bony defect is repaired with cartilage and/or soft tissue while leaving the function of the superior canal intact.
2. If cVEMPs were abnormal, why were VNG and RC results normal?
   VNG, RC, and cVEMP testing assess different segments of the vestibular system. VNG and RC testing assess vestibular

function by stimulating the lateral semicircular canals and neural links to eye muscles, pons, and cerebellar structures with rotational stimuli or caloric irrigation. cVEMP testing is specifically focused on the saccule and the neural connections with the sternocleidomastoid muscles and medulla.

3. Why are cVEMP thresholds lower (abnormal) in patients with SSCD, and why are air–bone gaps with normal bone conduction thresholds typically present on the audiogram?
   The measured cVEMP threshold is lower in patients with SSCD than in patients with no SSCD because the vestibular receptors become more sensitive to sound and pressure stimuli when the third window into the inner ear is created. A cVEMP stimulus presented to an ear with an SSCD results in increased saccular activation and in the cVEMP responses being present at lower than normal presentation levels. The third mobile window also allows for the dissipation of air-conducted acoustic energy/sounds, which causes a decrease in air-conduction thresholds. Bone-conducted thresholds are generally normal because the dehiscence does not alter or improve sound transmission within the bone.

32.8 Diagnosis and Recommended Treatment

32.8.1 Otologic Evaluation

The patient was examined by the neurotologist and was diagnosed with a dehiscence of the left superior semicircular canal. A discussion regarding the options of continued observation versus surgery to repair the SSCD occurred. The patient was advised regarding possible complications of surgery, including hearing loss/deafness, dizziness, vertigo, imbalance, facial paralysis, altered taste, infection, and bleeding. In spite of these concerns, the patient decided to go forward with surgery to repair the dehiscence.

A pre- and post-operative subjective questionnaire developed by the surgeon with questions regarding balance, hearing, and the overall global severity of symptoms was completed by KH (refer to “Key” in Table 32.1). The patient responded with a severity level (0–5, where a report of a 5 indicates symptoms that are most severe) for symptoms experienced in each category both pre-operatively and post-operatively (3 and 11 weeks). Post-operative responsees were compared with

Table 32.1 Subjective questionnaire of symptoms relating to semicircular canal dehiscence syndrome

	B1	B2	B3	B4	H1	H2	H3	H4	H5	GS1	GS2
Pre-op	5	5	4	5	3	3	5	5	0	5	5
3 weeks Post-op	2	1	0	5	1	2	5	1	0	3	3
11 weeks Post-op	0	1	0	0	0	3	5	0	0	1	1
Improvement											
KEY:											
Score: Use a “0” for “no symptoms.” Otherwise, rate the severity of the symptoms for each question.											
0: No symptoms											
1: Slight symptoms											
2: Mild symptoms											
3: Moderate symptoms											
4: Severe symptoms											
5: Profound symptoms											
Balance Symptoms:											
B1: Do you experience dizziness, vertigo, or imbalance?											
B2: Do you experience vertigo or imbalance caused by loud noises?											
B3: Do you experience vertigo or imbalance caused by changes in external pressure?											
B4: Do you experience vertigo or imbalance caused by coughing, sneezing, straining?											
Hearing Symptoms:											
H1: Do you have a sensation of fullness in one or both ears?											
H2: Do you have any trouble with your hearing?											
H3: Do you hear a ringing, buzzing, or humming noise in one or both ears?											
H4: When you speak or breathe, does it sound louder in one ear?											
H5: Do you hear your own heartbeat, eye movements, chewing, or footsteps?											
Global Severity Index:											
G1: Do your ear symptoms have a negative impact on your daily activities?											
G2: Overall, how would you rate the severity of your ear symptoms?											

pre-operative responses to determine if a change in outcome resulted. As seen in Table 32.1, pre-operatively, KH reported severe to profound symptoms of vertigo, dizziness, or imbalance triggered by loud noises, external pressure changes, coughing, sneezing, or straining. Additionally, he reported that the severity of hearing loss, aural fullness, tinnitus, and autophony in the affected ear ranged from moderate to profound in degree. The patient denied hearing internal body noises (heartbeat, eye movements, chewing). Overall, KH reported that his pre-operative auditory and vestibular symptoms had a profound negative impact on his daily activities and quality of life.

32.8.2 Surgery

A cartilage cap technique as described by Lundy et al was performed. A tragal cartilage graft was harvested. A cortical mastoidectomy was performed. The lateral, posterior, and superior semicircular canals were identified. A small area of bone (1 × 1 cm) of the tegmen (middle fossa floor) just lateral to the superior canal was removed using a diamond burr to expose the dura. Using a small elevator, the dura overlying the superior canal was elevated creating a small pocket. The cartilage graft was positioned above the superior semicircular canal within the pocket to reconstruct the dehiscence. The cartilage extended just lateral to the semicircular canal to support the area of dura that had been exposed during the procedure.

32.9 Outcome

As seen in Table 32.1, at 11 weeks following surgery to repair the SSCD, KH reported a significant improvement in dizziness and balance complaints. He denied any symptoms of vertigo, dizziness, or imbalance provoked by changes in external pressure, coughing, sneezing, or straining and reported only slight symptoms provoked by loud noises. Despite denying a reduction in the amount of tinnitus and hearing loss, he noted a slight improvement in aural fullness and autophony in the affected ear. Overall, KH reported that his auditory and vestibular symptoms improved significantly after the surgery to repair the SSCD and that his remaining symptoms only have a slight impact on his daily activities and quality of life.

32.10 Key Points

1. SSCD is caused by the absence of bone over the superior semicircular canal and can yield symptoms of pressure- and sound-induced vertigo and oscillopsia. Hearing related symptoms include hearing loss, pulsatile tinnitus, aural fullness, hyperacusis (oversensitivity to sound), and autophony in the affected ear(s).
2. SSCD is likely congenital, but slow bone erosion and head trauma are also thought to be indicated in the pathogenesis of the disorder.
3. Clinical indications of SSCD consist of air–bone gaps, generally low-frequency with normal or better than normal bone-conduction thresholds, normal tympanograms, present ARTs, cVEMP thresholds below 70 dB nHL, and CT scan of the temporal bone positive for SSCD on the affected side(s).
4. Counseling to understand the condition, avoidance of precipitating stimuli, and/or surgery are the treatment options for SSCD.

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33 Hyperventilation-Induced Nystagmus

Heather Monroe

A 59-year-old female with a right solitary vestibular schwannoma was seen for vestibular examination due to symptoms of persistent imbalance.

33.1 Clinical History and Description

GM is a 59-year-old female with a 10- to 15-year history of gradual bilateral progressive hearing loss. In June 2010 GM was hospitalized for several days for an acute episode of severe vertigo. Her balance was markedly impaired, and when she was discharged from the hospital she was using a walker to ambulate (i.e., walk). Prior to the vertiginous attack, GM began noticing increased difficulty hearing in her right ear. She was given prednisone but did not notice any improvement in her hearing. While GM was in the hospital, magnetic resonance imaging (MRI) was completed and was reported as “unremarkable.” Her balance gradually improved in the following months but never returned to “normal.” She was referred to physical therapy to address her chronic issues with balance but did not feel she received any benefit from the therapy. Over the next 2 years the hearing in her right ear rapidly declined, and she was fit with a left bilateral contralateral routing of the signal (BICROS) hearing aid.

In May 2012, GM was referred to a neurotologist to further investigate her symptoms of sudden hearing loss in her right ear and her persistent imbalance. GM described her symptoms as being off-balance, veering to the right and never feeling straight, lightheadedness, feeling like she is not “in contact” with her surroundings, and a constant feeling of being “hung over.” In addition, she reported fullness and constant tinnitus in her right ear. The neurotologist contemplated a number of possible diagnoses, including genetic hearing loss, endolymphatic hydrops, vestibular schwannoma, labyrinthitis, and autoimmune disease. Upon personal review of the MRI scan from 2010, the neurotologist identified a small vestibular schwannoma in the right internal auditory canal. A second MRI scan confirmed the presence of a tubular mass measuring 1 cm along the axis of the cranial nerve VII–VIII complex, 4 mm in the transverse dimension, and extending 7 mm into the internal auditory canal consistent with vestibular schwannoma. The neurotologist ordered an audiological examination, auditory brainstem response (ABR) testing, and vestibular examination.

33.2 Audiological Testing

The patient provided copies of her audiological examinations dating back to May 2011. Audiological examinations prior to that were not available. The patient reported that before her sudden hearing loss the right ear was her better-hearing ear and the ear she used on the telephone.

The audiological examination completed in May 2011 revealed significant asymmetry in word recognition scores (WRSs), which were 8%in the right ear and 100%in the left ear. Pure-tone thresholds revealed a mild sloping to moderate rising to mild sensorineural hearing loss in the right ear and a slight sloping to moderate rising to normal sensorineural hearing loss in the left ear (Fig. 33.1). Audiological examinations completed over the following months revealed a steady decline in pure-tone thresholds in the right ear. An audiological examination in September 2012 revealed a severe rising to moderately severe sensorineural hearing loss in the right ear with a WRS of 0%(Fig. 33.2).

GM’s ABR testing revealed normal absolute latencies for Waves I, III, and V on the left side and absent waveforms on the right side (Fig. 33.3). Increased stimulation repetition rate in the left ear revealed a normal shift in Wave V latency. Increased stimulation repetition rate was not tested in the right ear due to absent waveforms at the lower repetition rate. The results from the ABR test were consistent with retrocochlear pathology in the right ear.

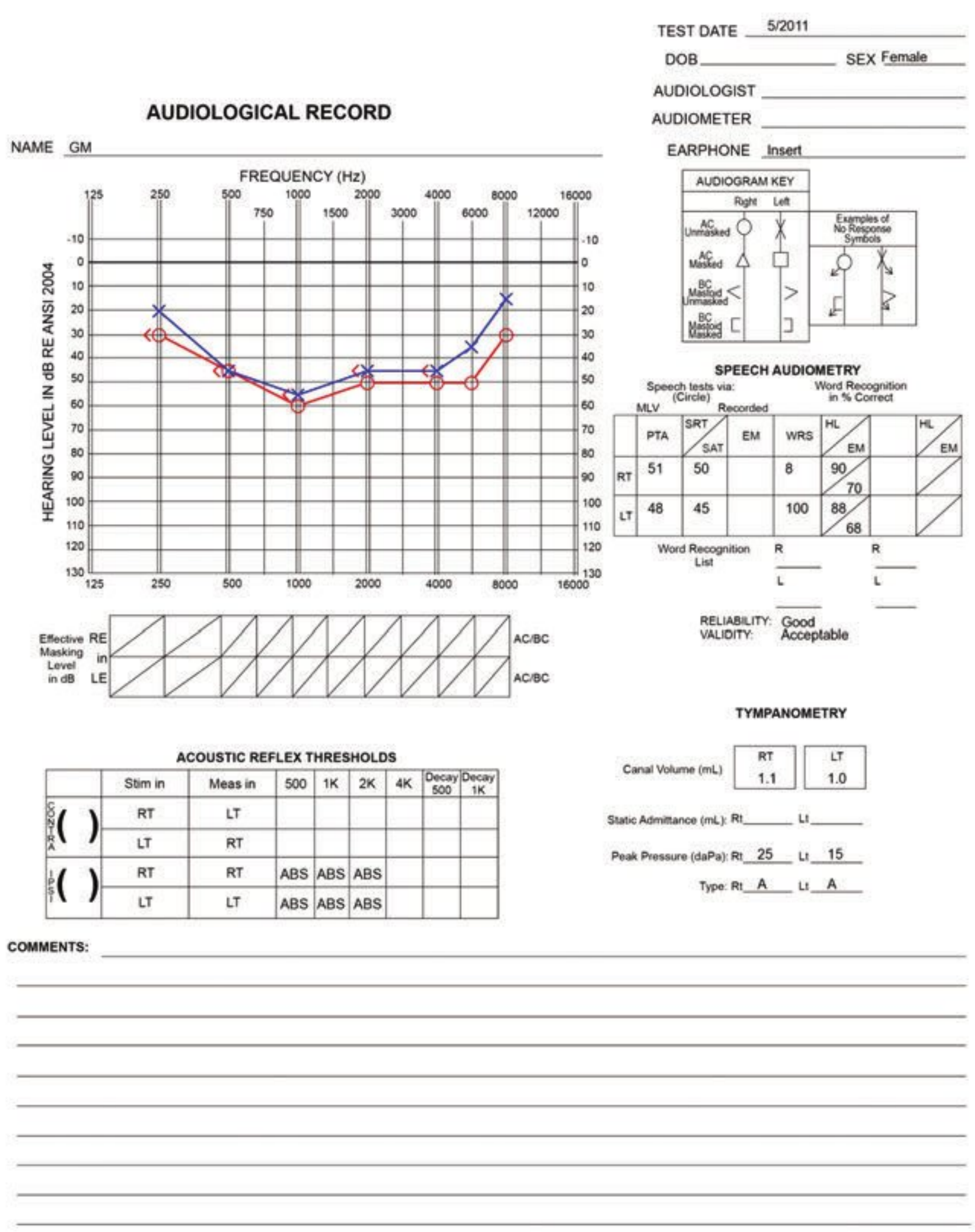
33.3 Vestibular Testing

The patient was seen for initial vestibular examination in June 2012. Video-oculography (VOG) revealed a 42% right reduced vestibular response. Computerized dynamic posturography (CDP) was normal with the exception of a backward center of gravity.

In November 2012 she returned for additional vestibular testing including CDP, cervical vestibular-evoked myogenic potentials (cVEMPs), and a rotational chair (RC) study. A decline in performance on CDP was observed when compared with previous testing. CDP revealed falls on the sensory organization test (SOT) 5 condition and some difficulty on the SOT 6 condition, indicating that effective use of vestibular input was poor. In addition, GM had difficulty with toes-down rotations of adaptation, and center of gravity remained backward. cVEMP responses were within normal limits with responses obtained and replicated to a threshold of 90 dB nHL in the left ear and 95 dB nHL in the right ear. Dynamic subjective visual vertical (SVV) testing revealed a 3° tilt to the right when rotating on axis, a 1° tilt to the left during eccentric rotation stimulating the right ear, and a 5° tilt to the right during eccentric rotation stimulating the left ear. Dynamic SVV results were consistent with a right unilateral vestibular weakness. The remainder of the RC study was unremarkable. Evaluation of hyperventilation was completed and revealed a 4°/s rightward-beating nystagmus after hyperventilating for approximately 20 seconds. The nystagmus increased to 8°/s rightward beating then gradually decayed until the nystagmus was no longer present. Hyperventilation nystagmus with the fast phase beating toward the weaker ear suggested that the lesion was excitatory. It was unclear whether the irritative focus was coming from the nerve or the ear.

33.4 Questions to the Reader

1. Do patients with vestibular schwannomas typically report symptoms of dizziness?



AUDIOLOGICAL RECORD

NAME GM

TEST DATE 5/2011
DOB ____ SEX Female
AUDIOLOGIST ____
AUDIOMETER ____
EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	51	50		8	90 / 70		
LT	48	45		100	88 / 68		

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.1	1.0

Static Admittance (mL): Rt____ Lt____
Peak Pressure (daPa): Rt 25 Lt 15
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT	ABS	ABS	ABS			
	LT	LT	ABS	ABS	ABS			

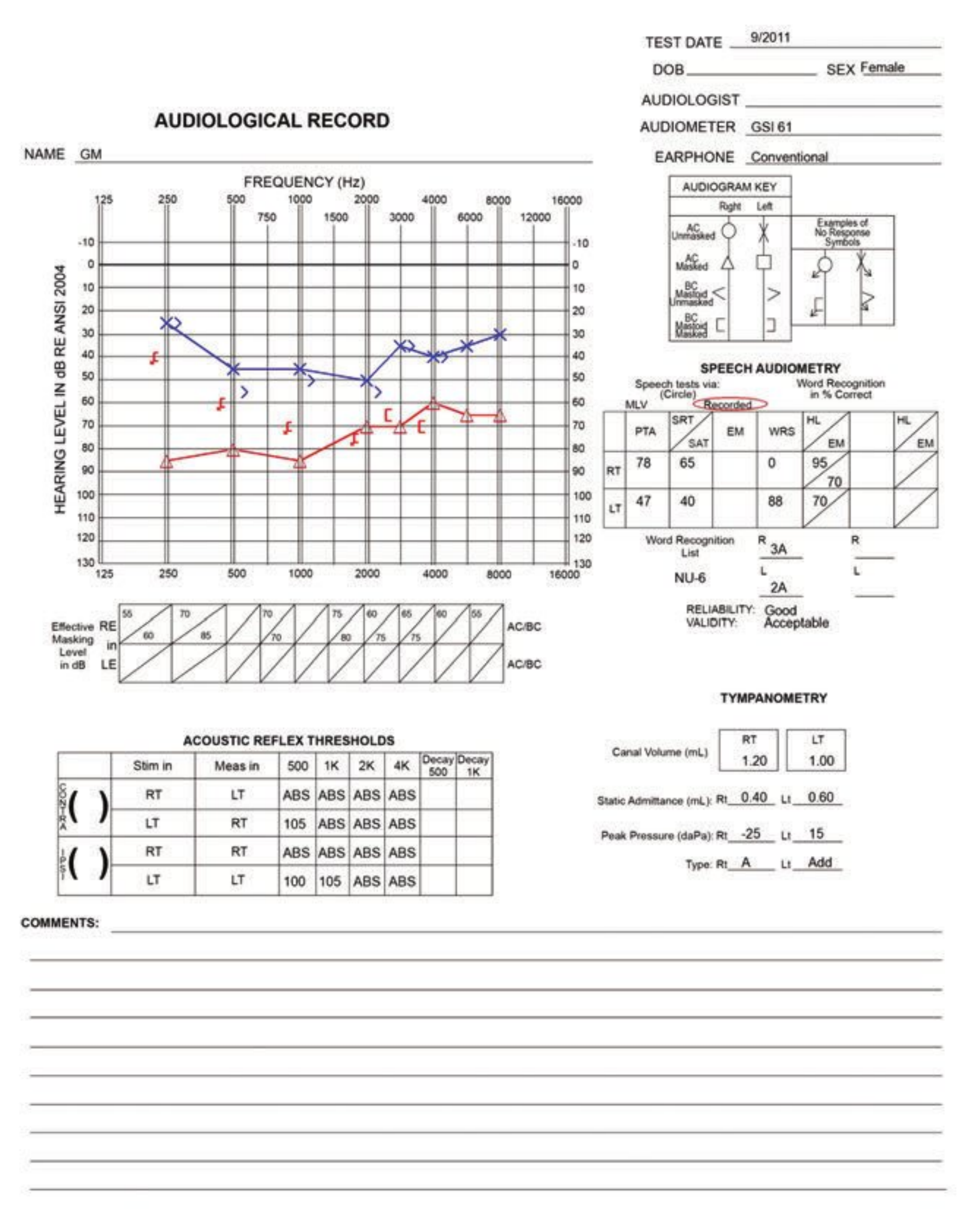
COMMENTS: ____

Fig. 33.1 Audiological examination from an outside facility completed May 2011. This examination was completed after the patient had noticed a significant change in the hearing in her right ear. Note the poor word recognition score in the right ear.

2. How is hyperventilation-induced nystagmus evaluated and what does it indicate?
3. What is the dynamic SVV test?

33.5 Discussion of Questions to the Reader

1. Do patients with vestibular schwannomas typically report symptoms of dizziness?
   Vestibular complaints are uncommon in patients with vestibular schwannoma due to the slow growth rate of the tumor. As the tumor increases in size, symptoms of dizziness and imbalance become more likely, but these symptoms are typically mild and short-lived. Hearing loss and tinnitus are often the first and only signs of the vestibular schwannoma. In this case report, the patient's symptoms were unexpected for the small size of GM's tumor.
2. How is hyperventilation-induced nystagmus evaluated and what does it indicate?
   Evaluation of hyperventilation-induced nystagmus should be considered in patients who: have suspected or confirmed retrocochlear pathology, become symptomatic during moments of stress, or describe an onset of symptoms during aerobic activities such as running or climbing stairs. This can be accomplished by recording the patient's eye movements during hyperventilation using VOG equipment. Typically, the patient is asked to hyperventilate for 30 seconds or longer, and the eyes are observed for the presence of nystagmus. Nystagmus that occurs either during or immediately after hyperventilation is consistent with central or peripheral vestibular disease. Hyperventilation nystagmus is most prevalent in patients with retrocochlear pathology but has also been observed in patients with Ménière disease, vestibular neuritis, labyrinthine fistula, cerebellar disease, migraine, multiple sclerosis, and other diseases. Evaluation of hyperventilation nystagmus can help to identify if a known lesion is excitatory or inhibitory and can "unmask" unilateral vestibular disease or cerebellar disease when the remainder of the vestibular examination is normal. If hyperventilation reproduces the patient's symptoms of dizziness but no nystagmus is present, anxiety-induced dizziness should be considered.



TEST DATE 9/2011

DOB ______ SEX Female

AUDIOLOGIST ______

AUDIOMETER GSI 61

EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME GM

Effective Masking Level in dB											
RE in	55 60	70 85		70 70		75 80	60 75	65 75	60	55	AC/BC
LE											AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	78	65		0	95 / 70		
LT	47	40		88	70		

Word Recognition List NU-6 R 3A L 2A R ___ L ___

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	105	ABS	ABS	ABS		
IPSI ( )	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	100	105	ABS	ABS		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.20	1.00

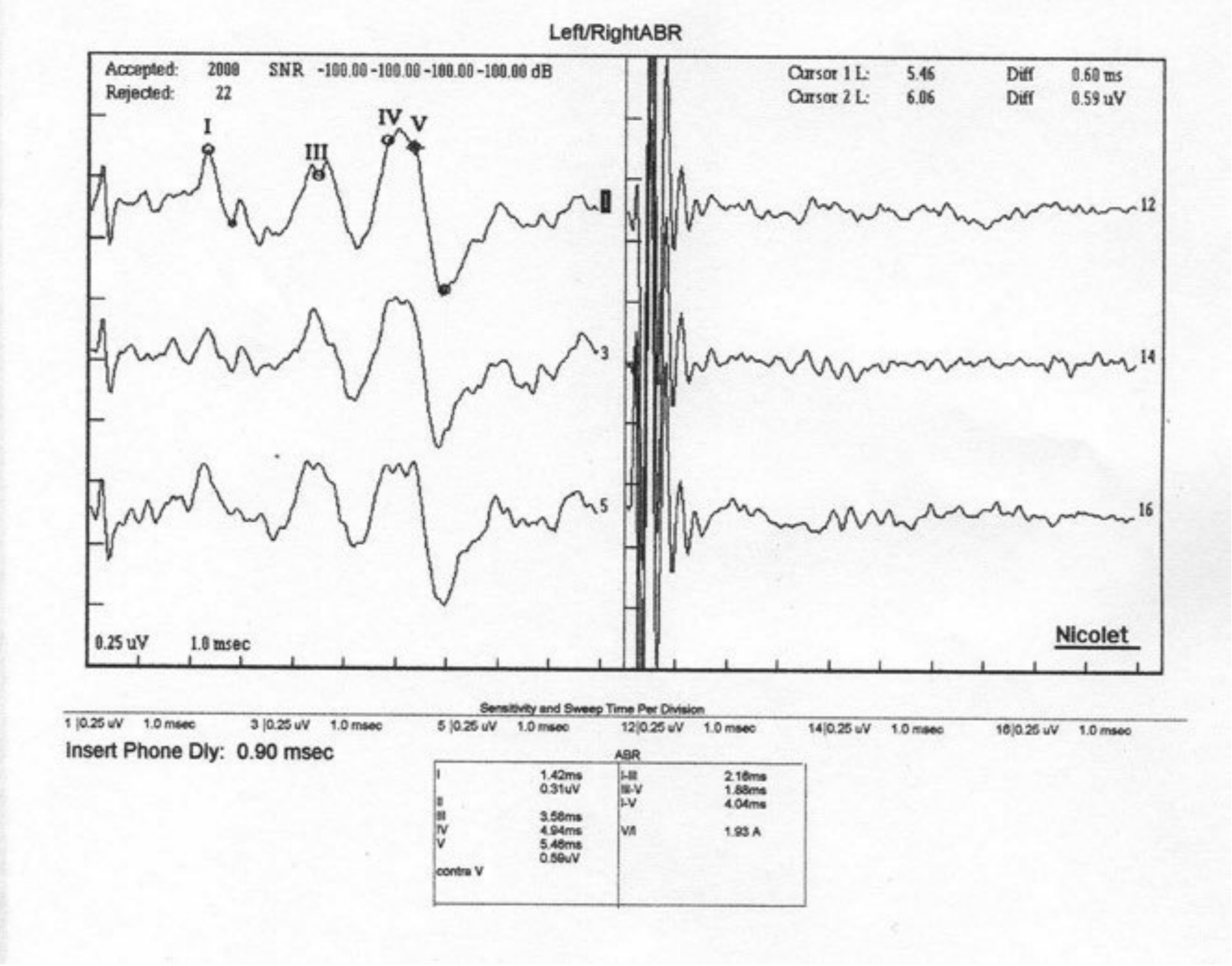
Static Admittance (mL): Rt 0.40 Lt 0.60

Peak Pressure (daPa): Rt -25 Lt 15

Type: Rt A Lt Add

COMMENTS:

Fig. 33.2 Audiological examination completed September 2012. Note the significant change in pure-tone thresholds in the right ear when compared with the audiological examination on May 2011.



Left/RightABR

Accepted: 2008 SNR -100.00 -100.00 -100.00 -100.00 dB
Rejected: 22

Cursor 1 L: 5.46 Diff 0.60 ms
Cursor 2 L: 6.06 Diff 0.59 uV

0.25 uV 1.0 msec

Sensitivity and Sweep Time Per Division

1 |0.25 uV 1.0 msec 3 |0.25 uV 1.0 msec 5 |0.25 uV 1.0 msec 12|0.25 uV 1.0 msec 14|0.25 uV 1.0 msec 16|0.25 uV 1.0 msec

Insert Phone Dly: 0.90 msec

ABR

I	1.42ms 0.31uV	I-III	2.16ms
		III-V	1.88ms
		I-V	4.04ms
II			
III	3.58ms		
IV	4.94ms	V/I	1.93 A
V	5.46ms 0.59uV		
contra V			

Fig. 33.3 Auditory brainstem response testing completed September 2012. Wave I through V latencies are normal in the left ear and absent in the right ear.

3. What is the dynamic SVV test?
   The dynamic SVV test is a measurement of utricular function. SVV is performed with the patient positioned upright in the RC and rotating at 300° per second in a light-proof enclosure to avoid visual reference. The patient is instructed to adjust a light bar so it appears earth-vertical before and during rotation of the chair and a difference in tilt between the two adjustments is recorded. This procedure is completed three times. First, the chair is rotated on axis so that both utricles receive equal stimulation. Next, the chair is positioned off axis and shifted approximately 4 cm to the left or right, so that the utricle positioned on axis receives minimal to no stimulation and responses to rotation come primarily from the eccentric utricle. In the case of two normally functioning utricles, minimal tilt is expected during on-axis rotation because responses from the two utricles cancel each other. When rotating off axis, a tilt away from the stimulated utricle is expected. In patients with unilateral utricular dysfunction, the examiner can expect to see a tilt toward the weaker ear when rotating on axis, minimal tilt when stimulating the weaker ear, and a tilt toward the weaker ear when stimulating the stronger ear.

33.6 Diagnosis and Recommended Treatment

The patient was diagnosed with a right solitary vestibular schwannoma and bilateral hereditary recessive sensorineural hearing loss. The patient was scheduled for gentamicin injections in the right ear in an effort to make the potentially fluxing excitatory lesion a stable inhibitory lesion and to help define locus. Translabyrinthine surgery to remove the tumor was to be considered if the gentamicin was ineffective in relieving her symptoms, although the neurotologist was concerned that surgery could make her symptoms worse.

33.7 Outcome

At her follow-up appointment following the second gentamicin injection the patient reported further decline in her balance. She reported difficulty in open spaces, climbing bleachers, walking and looking around, stumbling in the dark, and navigating narrow walkways such as walking between rows of chairs. VOG that day revealed an 8°/s leftward beating post-headshake nystagmus (beating toward the stronger ear), no change in the symmetry of the caloric response (although a decrease in total eye speed was noted), and a 12°/s rightward beating hyperventilation nystagmus. The physician prescribed amitriptyline to treat the patient's anxiety. Future physical therapy was discussed focusing on the patient's backward center of gravity and self-described highly visually weighted sense of balance.

33.8 Key Points

1. The presence of hyperventilation nystagmus can provide clarity in determining if a vestibular schwannoma is an excitatory or inhibitory lesion. Hyperventilation nystagmus with a fast component that beats toward the side of the vestibular schwannoma indicates that the lesion is excitatory.
2. The onset of symptoms during hyperventilation when no nystagmus is present may indicate anxiety-induced dizziness.
3. The vestibular apparatus consists of five matched pairs of acceleration-sensing organs. Most vestibular tests focus on the function of just one of these organs: the horizontal semicircular canal. It is possible, therefore, for vestibular tests to be normal despite the presence of vestibular disease. In some instances, hyperventilation nystagmus may provide the only evidence of unilateral vestibular disease or cerebellar disease.

33.9 Suggested References

Bance ML, O'Driscoll M, Patel N, Ramsden RT. Vestibular disease unmasked by hyperventilation. Laryngoscope 1998; 108: 610–614

Califano L, Melillo MG, Vassallo A, Mazzone S. Hyperventilation-induced nystagmus in a large series of vestibular patients. Acta Otorhinolaryngol Ital 2011; 31: 17–26

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34 Right Internuclear Ophthalmoplegia following a Stroke

Heather Monroe

A 58-year-old male with persistent imbalance was seen for vestibular examination following a right pontine angle stroke.

34.1 Clinical History and Description

MF is a 58-year-old male admitted to the hospital with symptoms of imbalance, dizziness, diplopia, nausea, and vomiting. At admission, his blood pressure was 190/101 mm Hg. The patient reported horizontal and vertical separation of objects both at a distance and within a near field. The diplopia was more pronounced with left gaze. Prior to arriving at the hospital he experienced tingling on the left side of his face, around his mouth, and in his left hand distally. Previous medical history included hypertension, insulin-dependent diabetes, and hyperlipidemia. Magnetic resonance imaging (MRI) revealed a small infarct in the posterior pons on the right side with atherosclerotic disease throughout the posterior circulation. Once he was deemed stable, he was transferred to an acute care facility for rehabilitation and subsequently discharged.

One month after the stroke, the patient was evaluated by neuro-ophthalmology. A motility exam by the neuro-ophthalmologist revealed a right internuclear ophthalmoplegia (INO). INO occurs in lesions of the medial longitudinal fasciculus (MLF) and appears as paresis of the ipsilateral eye as it moves inward toward the nose. Skew deviation, a condition where the eyes are vertically misaligned, was also noted. By the time of his follow-up appointment 1 month later, MF's diplopia had improved to the point where he had only minor difficulty when driving or reading. The right INO and skew deviation were reported by the physician as almost totally resolved.

MF was seen in our dizziness and balance laboratory 11 months after his stroke due to his persistent imbalance. At that time, the patient reported frustration with his sense of balance and his inability to move his head rapidly or process complex visual environments. He was not able to adequately perform his job in construction. When using power tools and machinery around his house he reported he had to concentrate more on each task. He reported a spinning sensation at times where the room appeared to move in a counterclockwise direction. In addition, he had difficulty moving his eyes rapidly back and forth.

34.2 Vestibular Testing

Air conducted cervical vestibular-evoked myogenic potential (cVEMP) responses were absent in both ears. A 4°/s leftward beating gaze nystagmus was present for leftward gaze without fixation in the left (abducting) eye only (Fig. 34.1). Saccades were disconjugate and consistent with a right INO. When the eyes were moving leftward there was a lag in the right (adducting) eye (Fig. 34.2). Bithermal caloric irrigation revealed a 48% left reduced vestibular response (Fig. 34.3). The remainder of the video-oculography (VOG) test was normal. Computerized dynamic posturography and rotational chair tests were also normal.

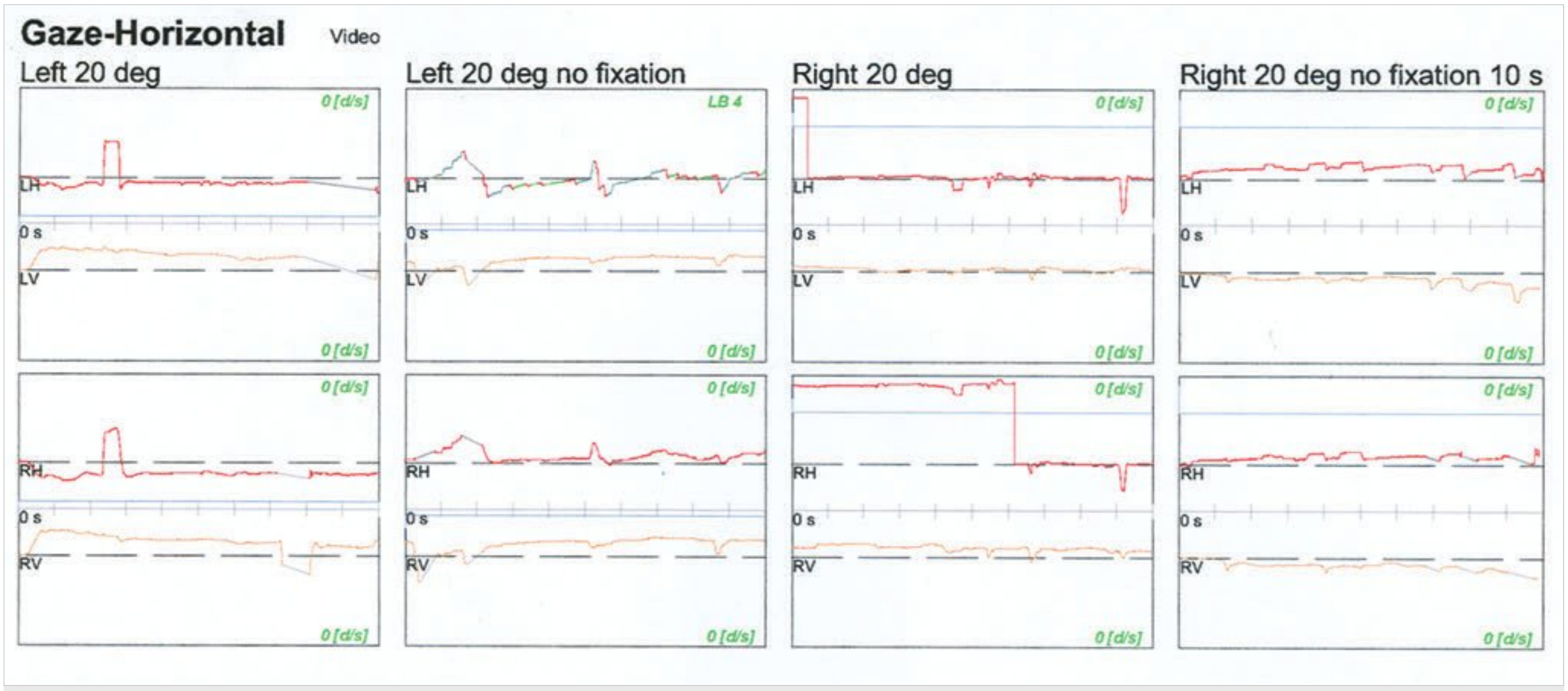


Fig. 34.1 Evaluation of gaze is represented with the eyes fixated on a target positioned 20° eccentric. In this figure, gaze was recorded first to the left then to the right. Gaze nystagmus is first measured with a target to fixate upon and then without fixation. The horizontal and vertical tracings for the left eye (LH and LV) are represented in the uppermost boxes, whereas tracings for the right eye can be found below (RH and RV). Note the 4° leftward beating nystagmus in the left eye when the patient gazes to the left without fixation.

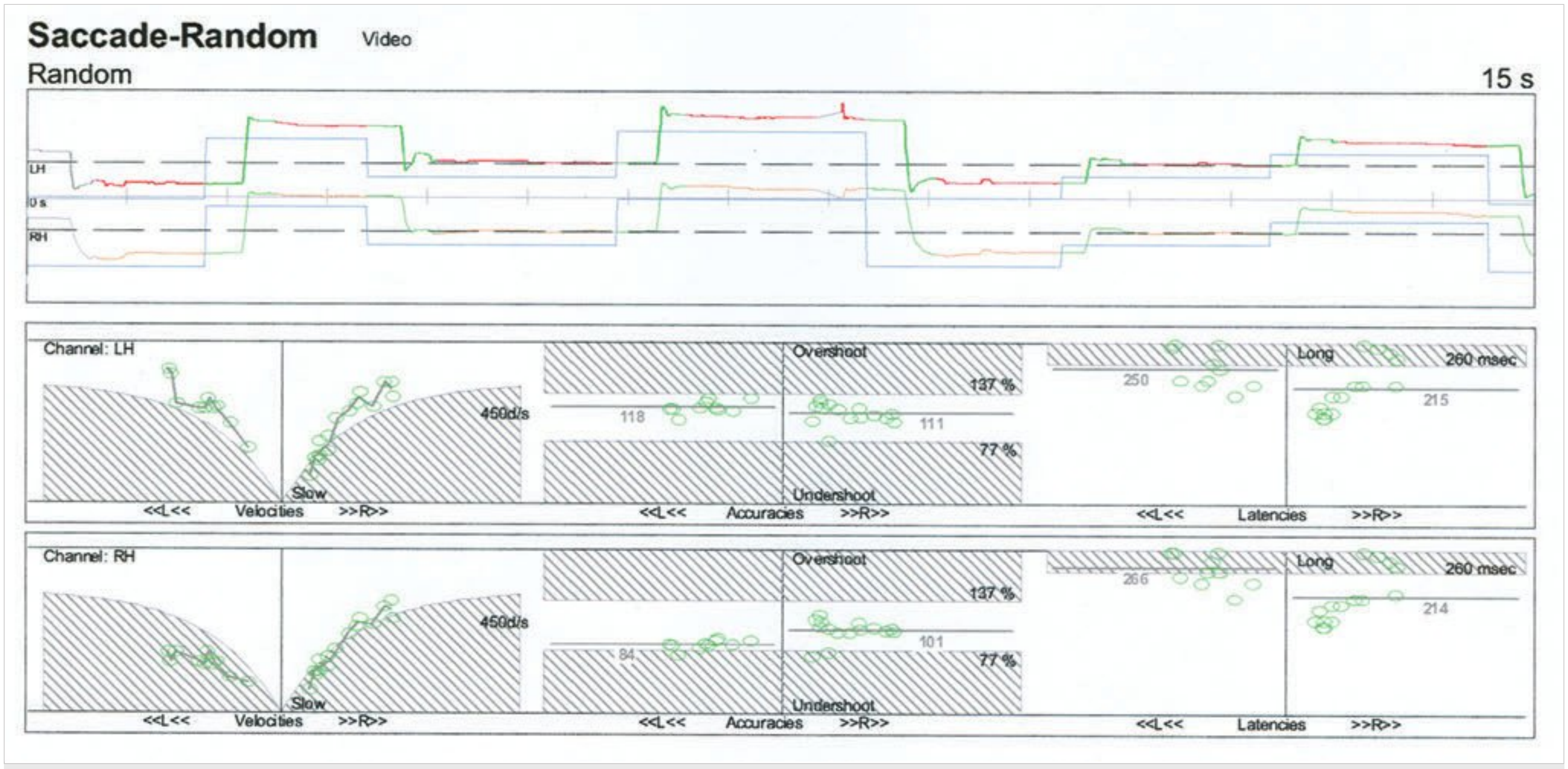
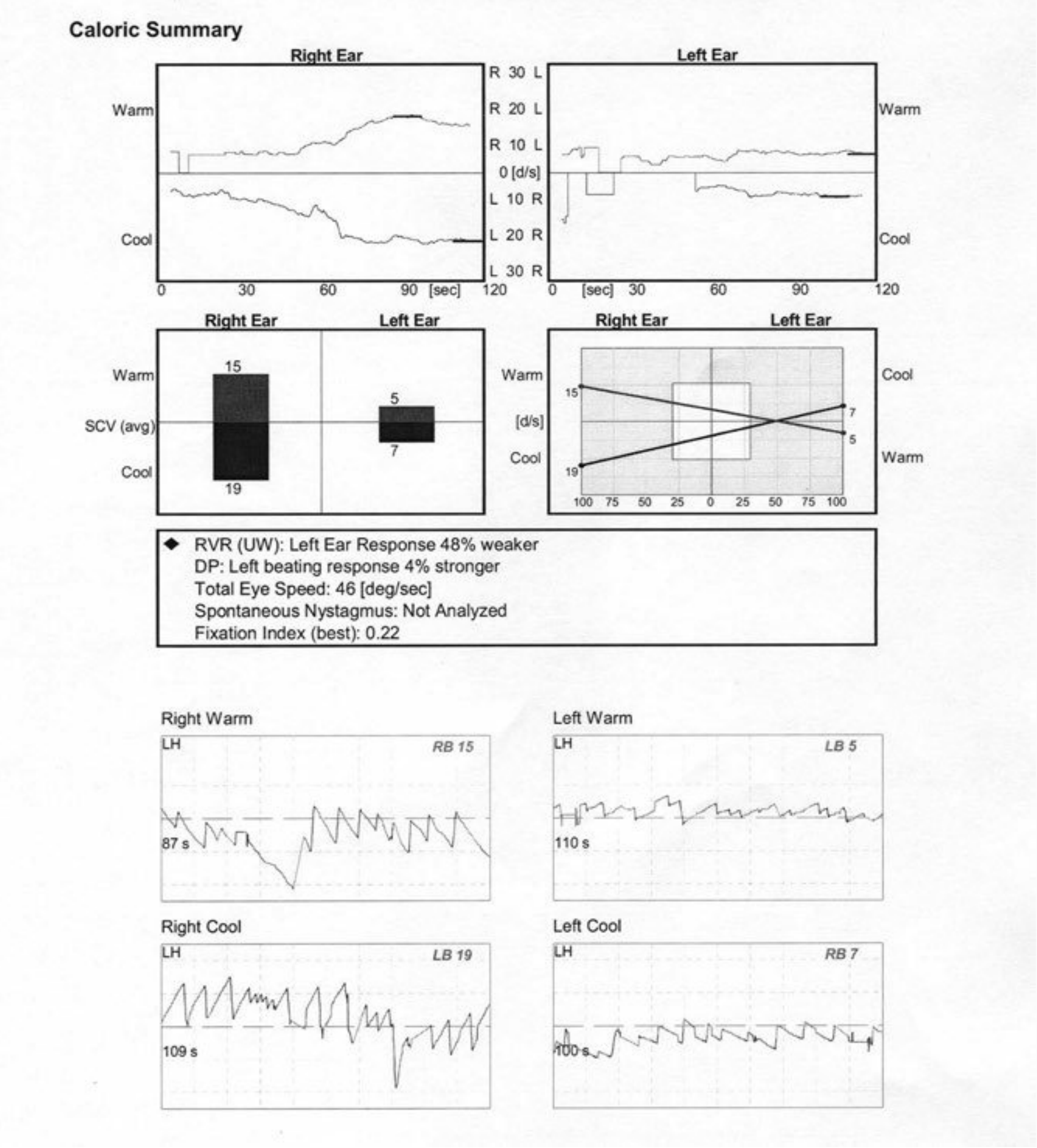


Fig. 34.2 Evaluation of random saccades is represented with tracings for the left eye on the uppermost graph and tracings for the right eye below. When looking at the graphs representing velocity and latency, there is a noticeable lag when the right eye was moving leftward.



◆ RVR (UW): Left Ear Response 48% weaker
DP: Left beating response 4% stronger
Total Eye Speed: 46 [deg/sec]
Spontaneous Nystagmus: Not Analyzed
Fixation Index (best): 0.22

Fig. 34.3 The caloric summary for bithermal caloric irrigations is displayed indicating a 48% left reduced vestibular response. The topmost boxes depict the progression of the response over 120 seconds with no nystagmus present at the beginning of the irrigation and then rising to a peak. The bar graph represents the peak response of each caloric irrigation. The intersection of the caloric responses displayed in the graph on the bottom right illustrates that the percentage difference between ears falls outside of the normative values for this laboratory (white box in the center). Calculations of the reduced vestibular response (RVR) or unilateral weakness (UW), directional preponderance (DP), total eye speed of all four irrigations, spontaneous nystagmus (in this case report no spontaneous nystagmus was present prior to irrigation), and fixation index are also visible.

34.3 Questions to the Reader

1. Why is the persistent INO a concern for balance?
2. What are some possible reasons for the absent cVEMP responses?
3. What parts of the VOG investigate central mechanisms?

34.4 Discussion of Questions to the Reader

1. Why is the persistent INO a concern for balance?

   Maintaining equilibrium is a complex task that involves the coordination of input from vestibular, visual, and somatosensory/proprioception systems. The vestibular and visual systems are interconnected through the vestibular ocular reflex, the function of which is to keep images stable on the retina during head movement.

   When the head is in motion, the eyes must be able to rapidly reposition in order to maintain objects of interest on the fovea. This rapid repositioning of the eyes is known as saccadic eye movement. When the eyes fail to move appropriately with head movement, images of the visual world "slip" on the retina producing blurred vision. The result is an inability to clearly see objects in the environment and determine their distance, size, and orientation. When walking, clear visualization of objects is necessary because it allows us to modify our path and avoid tripping or colliding with an object. When driving, gaze is quickly diverted to read road signs or to identify potential hazards in the road.

   In a patient with an INO, saccadic eye movements are disconjugate. In this particular case, when the patient looks to the left the right adducting eye lags behind. Therefore, when the patient moves his head or looks quickly from one object to the next, he is presented with a distorted visual image. This can explain why the patient complains of imbalance when moving his head rapidly and in complex visual environments.
2. What are some possible reasons for the absent cVEMP responses?

   cVEMP responses can be absent for a number of reasons. Studies have shown unilaterally decreased amplitude and absent cVEMP responses in patients with brainstem lesions, particularly if the lesion is in the lower brainstem. Amplitude is known to decrease with age, and by the sixth decade of life, cVEMP responses can be absent in normally functioning patients. Absent cVEMP responses may be the result of weak neck muscles, muscle stiffness, and inadequate muscle contraction. Mass and tone of the sternocleidomastoid (SCM) muscle not only vary between individuals but may also vary within the same individual, and therefore may be the cause of a unilaterally absent cVEMP response. Although a conductive hearing loss was not present in this patient, the reader is reminded that the air conducted cVEMP response can be abolished by even a mild conductive hearing loss due to the high intensity required to elicit a response. Vestibular end-organ pathologies such as Ménière disease and vestibular schwannoma may result in an absent cVEMP response, but typically the response is absent only in one ear. In addition, cVEMP responses may be absent in cases of idiopathic bilateral vestibulopathy.
3. What parts of the VOG investigate central mechanisms?

   The differentiation of symptoms caused by a peripheral lesion from those of central origin can be a difficult task. Abnormalities detected on tests of ocular-motor function can be an indication of central pathology when factors such as age, visual deficits, vigilance, alcohol/medication effects, drowsiness, and congenital abnormalities are taken into account. The ocular-motor function test battery on the VOG typically consists of gaze nystagmus, saccades, smooth pursuit, and optokinetics.

   Presence of gaze-evoked nystagmus is considered a central finding. In this case report, a leftward beating gaze nystagmus was present without fixation in the left eye only. Nystagmus of peripheral origin, however, may also be evident during gaze testing. Peripheral nystagmus is direction fixed, and follows Alexander's law, which states that nystagmus of peripheral origin becomes more intense with gaze in the direction of the fast phase and decreases with gaze in the direction of the slow phase.

   Analyses of saccade testing evaluate accuracy, latency, and velocity measures. In this case report, the saccades were disconjugate when looking to the left with the right eye lagging behind the left, consistent with a right INO. Other possible signs of central dysfunction that can be identified on saccade testing include saccades that inaccurately meet the target or demonstrate prolonged latencies. Saccadic dysmetria can occur in cerebellar disease, cerebellar pontine angle pathology, or brainstem lesions. Saccadic overshoots may be present in an INO, ipsilateral to the MLF lesion. It should be noted that inaccurate saccades may be attributed to visual deficits or muscle/nerve weakness. Prolonged saccades are a central finding but may also be due to inattention, medication effects, or drowsiness.

   Smooth pursuit is a test where the patient is instructed to smoothly track a predictable target as it moves in the horizontal or vertical plane. Smooth pursuit is sensitive to central pathology, but localization is difficult due to the complexity and redundancy of the system. In this case report, smooth pursuit remained intact. Analyses of smooth pursuit include measures of gain, symmetry, and phase. Gain measures reveal how accurately the patient was able to move his or her eyes with the target. Saccadic corrections can be seen when the eyes are unable to keep up with the target and are considered a central finding. Asymmetry on the tracing occurs when there is a significant difference in the gain when the eye is moving rightward versus leftward. Phase is an indication of how much the eyes are lagging or leading the target and may indicate the need for reinstruction. It should be noted that a progressive increase in the amount of saccadic corrections and the inability to smoothly track a target has been shown to occur with age.

   Optokinetic nystagmus is a test of ocular-motor function and should be performed with a stimulus that fills 90% of the visual field for adequate retinal stimulation. In our laboratory, optokinetic testing is performed during rotational chair testing where vertical lines are projected on the walls of the 360° enclosure surrounding the patient. In this case report optokinetic nystagmus was within normal limits. Asymmetry in optokinetic nystagmus may indicate a central lesion. Ocular-motor function tests are a good tool for evaluating central mechanisms. One should, however, be mindful that central mechanisms can have an impact on all aspects of the VOG. In this case report, an asymmetry was noted in the caloric response. Due to the lack of having access to previous vestibular testing, it is difficult to determine if the 48%left reduced vestibular response (Fig. 34.3) is a result of the stroke, or a previous peripheral insult. A spontaneous nystagmus is considered a central finding when it beats in a vertical direction, is enhanced with fixation, or is direction changing. Positional nystagmus is nonlocalizing and can be present in both peripheral and central lesions. Post-headshake nystagmus is considered a central finding if it beats in a vertical direction. Lack of fixation suppression following caloric irrigation is also a central finding. For a more in-depth review of central mechanisms and the interpretation of ocular-motor function tests the reader is encouraged to study one or more of the excellent resources listed in the reference section at the end of this case report.

34.5 Diagnosis and Recommended Treatment

The patient was diagnosed with post–right pontine angle stroke with a persistent right INO and imbalance. Distortion with rapid head movement or changes in gaze from the persistent INO were found to be the most significant concerns. At the last appointment with the patient's neurotologist he was instructed to continue physical activities as much as possible and minimize complex visual environments, which require him to perform rapid saccades accurately. This would include situations such as walking through a crowded marketplace where one must pay attention to the movement of other people in the crowd, read signs, and inspect objects for sale.

34.6 Outcome

The patient followed up with his neurologist 18 months after the stroke. At that time he reported continued diplopia and imbalance. In addition, he was experiencing significant post-stroke depression and insomnia. At the time of this writing the patient had not been seen for follow-up with his neurotologist or neuro-ophthalmologist.

34.7 Key Points

1. INO is a condition seen in lesions affecting the medial longitudinal fasciculus including stroke.
2. The visual disturbance associated with INO can have a significant impact on equilibrium and the patient's quality of life.
3. Collaborative teamwork and appropriate referral are essential to proper patient management. Although the neuro-ophthalmologist felt that the INO was almost completely resolved, it was still a major concern from the perspective of the neurotologist.
4. An accurate case history is a vital component in the diagnosis and management of the dizziness- and balance-disordered patient. It can be the key to differentiating between peripheral and central findings, indicate what testing is appropriate, and help determine the course of treatment.

Suggested Reading

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35 Unilateral Utricular Dysfunction Presenting as Disorientation while Driving

Belinda C. Sinks

This case report summarizes a patient with unilateral utricular dysfunction presenting itself as an isolated symptom of dizziness only during driving.

35.1 Clinical History and Description

TL, a pleasant 43-year-old female presented with a 10-month history of a very specific sensation of tilting only during high velocity turns and only when veering leftward. She had no precipitating episode of vertigo, nausea, vomiting, nor any tinnitus or change in hearing. She stated she had to pull over while driving because she felt she would roll the car or fly off the highway. TL also reported she had tried diazepam and meclizine neither of which was beneficial. TL stated she was at a point where she was not comfortable driving on highways requiring higher speeds and these specific kinds of turns. TL presented with results of previous tests that included magnetic resonance imaging (MRI), lab work, audiometry, auditory brainstem response (ABR), and videonystagmography (VNG).

Results from the MRI with and without contrast were reviewed. Without contrast, findings suggested no acute infarcts as demonstrated by diffusion-weighted imaging. With contrast, several nonspecific focal areas of T2 signal alteration in the subcortical bilateral cerebral hemispheres were noted. Differential diagnoses from the MRI included migraine headache, demyelinating process, or small vessel disease. Infectious or inflammatory processes would have a similar appearance but are statistically less likely. Low-lying cerebellar tonsils were also noted, although an Arnold-Chiari malformation was excluded upon later review. Complete blood count (CBC), comprehensive metabolic panel, thyroid panel, and sedimentation rate studies were all within normal limits.

The audiological examination revealed normal hearing at 250 to 8000 Hz, bilaterally. Speech recognition thresholds (SRTs) agreed with the pure-tone average (PTA) and revealed normal ability to receive speech bilaterally. Word recognition scores (WRSs) were 100%bilaterally, indicating normal ability to recognize speech. Auditory brainstem response (ABR) testing revealed normal absolute latencies and interpeak intervals, bilaterally.

VNG revealed no spontaneous nor gaze nystagmus with or without fixation. Horizontal tracking and horizontal saccade results were within normal limits. Optokinetic nystagmus was symmetric and within normal limits. Dix-Hallpike maneuvers were negative bilaterally. Bithermal caloric irrigations were within normal limits with a 22%right relative reduced response. Total eye speed was not available. Fixation suppression was successful, and there was no directional preponderance.

TL was referred for additional evaluation and results of the physical examination by a neurotologist are as follows. Vital signs revealed a blood pressure of 132/80 with a heart rate of 60 beats/min. Her tympanic membranes were normal and mobile. Her air conduction sensitivity was greater than bone conduction bilaterally without lateralization using a 512 Hz tuning fork. TL demonstrated no spontaneous or gaze-induced nystagmus. She had normal saccades and smooth pursuit. Head impulse testing was negative as was post-headshake testing. Dix-Hallpike responses were negative bilaterally. TL exhibited excellent stability on a firm surface and on a 3-inch foam pad with her eyes closed. Her gait was normal. TL was referred for cervical vestibular-evoked myogenic potential (cVEMP) and rotational chair (RC) testing to include dynamic subjective visual vertical (DSVV). This was completed immediately following her physical exam.

35.2 Vestibular Testing

cVEMP testing was completed using rarefaction 500 Hz tone burst stimuli presented through insert earphones. Waveforms were obtained and replicated at an intensity level of 95 dB nHL, bilaterally. Thresholds were obtained at a level of 90 dB nHL bilaterally. These results imply bilateral integrity of the saccule and sacculocollic pathways.

RC testing revealed slightly decreased vestibulo-ocular reflex (VOR) gain with normal symmetry and phase. Step velocity time constants were within normal limits for the per- and post-rotary chair conditions. Optokinetic nystagmus was symmetric and within normal limits. Visual-vestibular fixation suppression was intact.

DSVV testing revealed a tilt of 3° to the right during centric rotation (□ Fig. 35.1). Eccentric rotation when stimulating the right utricle resulted in no change in tilt (□ Fig. 35.1). A significantly low deviation of DSVV implies abnormal processing of otolith information, which is predominantly related to utricular input. Eccentric rotation when stimulating the left utricle resulted in an 8° degree tilt to the right (□ Fig. 35.1).

35.3 Questions to the Reader

1. How was the DSVV performed?
2. Was DSVV testing helpful in diagnosing vestibular dysfunction in TL?
3. What vestibular tests provide information regarding the otolith system?
4. Why did TL have symptoms only when driving on leftward curves?
5. What are the limitations of DSVV?

35.4 Discussion of Questions to the Reader

1. How was the DSVV performed?
   This test was performed with a vertical axis RC with a five-point safety belt. The head was stabilized with a head rest and cushioned Velcro strap preventing any movement. A

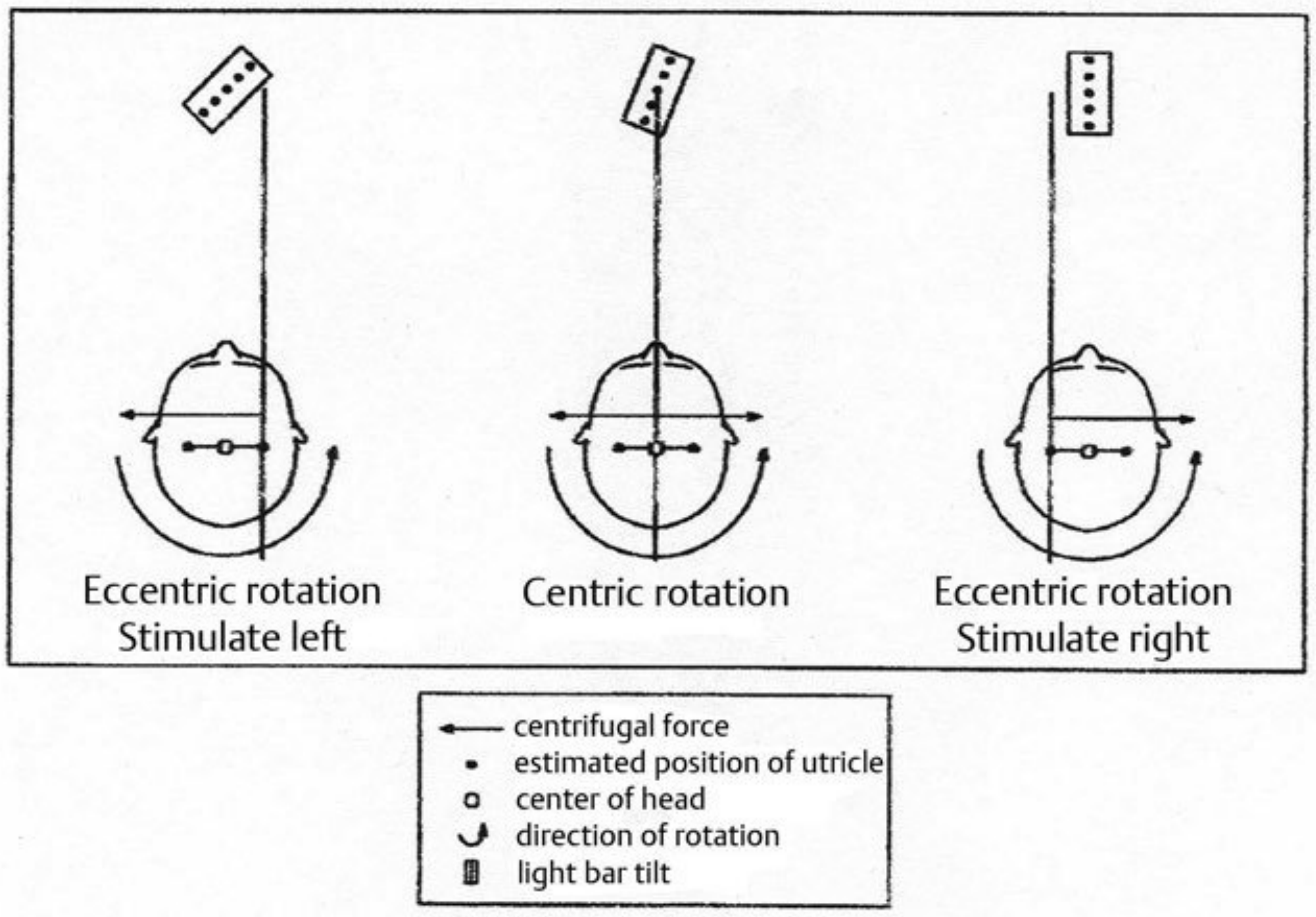

Fig. 35.1 Representation of the light bar tilt with eccentric rotation stimulating the left ear. There was an 8° change in tilt to the right. This result is consistent with normal function. Next, representation of the light bar tilt with centric rotation. There was a 3° tilt to the right. Lastly, representation of the light bar tilt with eccentric rotation stimulating the right ear. There was no change in tilt. This suggests an abnormality in the system.

light bar was secured at eye level in front of TL. Prior to testing, the room was darkened and TL was asked to adjust the light bar to vertical. The alignment of the light bar (in degree) was recorded via an infrared camera. TL was instructed to randomly tilt the light bar off center and to close her eyes. The chair was accelerated on center rotation at 10°/s^2 to a velocity rate of 300°/s. When the chair was at 300°/s for 60 seconds, TL was instructed to open her eyes and realign the light bar to vertical once again. The chair was then decelerated at 5°/s^2 to 0/s.

The procedure was repeated when the chair was translated 4 cm lateral in the coronal plane to eccentric position to the right and then to the left. Translating the patient laterally along the interaural axis at the half of interutricular distance approximately 4 cm, this technique allows a vertical axis rotation to pass through one utricle, which will be minimally stimulated while exposing the contralateral utricle to centrifugal acceleration at constant velocity. Duration of the entire test is approximately 10 minutes. The changes of alignment of the light bar were recorded (in degrees) separately in the centric as well as right and left eccentric rotations.

2. Was DSVV testing helpful in diagnosing vestibular dysfunction in TL?
   Based upon TL's history of only high-velocity stimulation causing her symptoms, RC testing with DSVV was helpful. The DSVV test allowed testing of each utricle separately, providing a comparison between each side as well as to static tilt. Subjective visual vertical tilt during centric rotation hinted of a slight tilt to the right. With eccentric rotation, however, putting the left ear "on axis" and stimulating only the right utricle, there was no change in tilt, which is abnormal. Placing the left ear into the stimulated position and the right ear on axis resulted in an approximate 8° of tilt as compared with static measures. The response from the left ear is highly significant and is more consistent with normal function than the response from the opposite ear.
3. What vestibular tests provide information regarding the otolith system?
   The DSVV test provides clinicians with important information regarding utricular function of each ear independently. Through centrifugation, the patient is stimulated via centric (on axis) rotation, which stimulates both ears, and eccentric (off axis) rotation stimulating one ear at a time. Ongoing research with ocular evoked myogenic potentials (oVEMPs) suggests that there are responses related to utricular origin. The cVEMP test provides clinicians with information regarding saccular function and the subsequent pathway. Computerized dynamic posturography (CDP) is the longest-standing indirect test of the effects the otolith has upon the control of balance.
4. Why did TL have symptoms only when driving on leftward curves?
   When TL was going around leftward curves at high speeds (60 mph) her outermost (right) utricle was being stimulated more than her left. In this scenario, the outermost utricle contributes the most gravitational information. TL's right utricle was not contributing appropriately to the sense of proper tilt; thus TL felt as if she were going to fly off of the highway.
5. What are the limitations of DSVV?
   Although DSVV testing relies heavily on utricular detection of centrifugation and gravity combined, one must recognize that the estimation of verticality also depends on other sensory inputs such as vision, proprioception, higher midbrain, and cortical spatial mapping. Therefore, the degree of DSVV tilt may not necessarily correspond with the degree of ocular counter-roll. There is an inability to effectively eliminate haptic cues, there are adaptive changes in otolith tone after injury, and there is significant deviation required to suggest impaired otolith tone (>2.5°). The literature does support, however, that DSVV deviation during eccentric rotation is a reliable measurement for identifying and localizing utricular asymmetry.

35.5 Diagnosis and Recommended Treatment

TL's test results suggested right utricular dysfunction. Habituation driving exercises were planned.

35.6 Outcome

TL participated in habituation driving exercises in a safe environment and limited her highway speed to a stable threshold. TL reported she had improved significantly through repeated stimulating maneuvers with a driving instructor.

35.7 Key Points

1. Taking a comprehensive case history, thorough medical exam, and diagnostic battery are all essential to the diagnosis and management of the balance-disordered patient.
2. Further investigations into otolith function and assessment are increasing in clinical and research laboratories around the world so clinicians can have better tools to assess and help these patients.

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36 Unilateral Peripheral Vestibular Dysfunction Confounded by Central Inhibition

Belinda C. Sinks

This case report summarizes a patient with unilateral dysfunction presenting itself as bilateral dysfunction.

36.1 Clinical History and Description

PD is a pleasant 39-year-old male with a 6-year history of occasional vertigo as well as imbalance with fast head or body turns. He had sought several medical opinions in the past and had been diagnosed with bilateral peripheral vestibular loss, bilateral perilymphatic fistulae, as well as benign positional vertigo. Past medical treatments included a repositioning maneuver for benign positional vertigo, sequential middle ear explorations, and subsequent packing for a suspected perilymphatic fistula. He then presented at our audiology clinic for further evaluation.

PD was seen by a neurotologist for his symptoms of persistent imbalance and vertigo. He stated that the world "jiggled" when he chewed or used handheld power tools that vibrated, and he indicated difficulty walking in the dark. Medical examination revealed intact tympanic membranes; normal nasal, oral, laryngeal, and neck exam; absence of spontaneous nystagmus; absence of gaze-evoked nystagmus; normal saccades and smooth pursuit; and negative Dix-Hallpike testing. He revealed a positive head impulse sign during head impulse testing (HIT) in the right horizontal plane. This meant that when the examiner moved PD's head quickly to the right in the horizontal plane, PD was unable to maintain fixation on the target and a refixation saccade was visible to the examiner. Dynamic visual acuity testing (DVAT) revealed markedly reduced results with performance diminishing more than seven lines on the Snellen eye chart. The DVAT test looked at the difference in visual acuity between PD's static visual acuity and when the examiner shook his head side to side. A decrease of greater than two lines on the chart is considered abnormal. Increased sway was noted while he stood with eyes closed. Also, he fell with delayed posture response when standing on 3-inch foam with his eyes closed. Magnetic resonance imaging (MRI) and computed tomographic (CT) scans were unremarkable. The medical impression was suspected bilateral vestibular dysfunction with idiopathic etiology. PD's physician ordered bithermal caloric irrigation, ice water caloric irrigation, cervical vestibular-evoked myogenic potentials (cVEMPs), and rotational chair (RC) studies.

36.2 Vestibular Testing

Bithermal caloric irrigation results were bilaterally reduced with a total eye speed of 12°/s. Ice water caloric irrigation revealed a response of 3°/s from the right ear and a 37°/s response from the left ear (Fig. 36.1). RC testing revealed decreased low-frequency vestibulo-ocular reflex (VOR) gain at 0.025 and 0.05 Hz with phase lead. There was an asymmetry with clockwise (rightward) rotations being weaker than

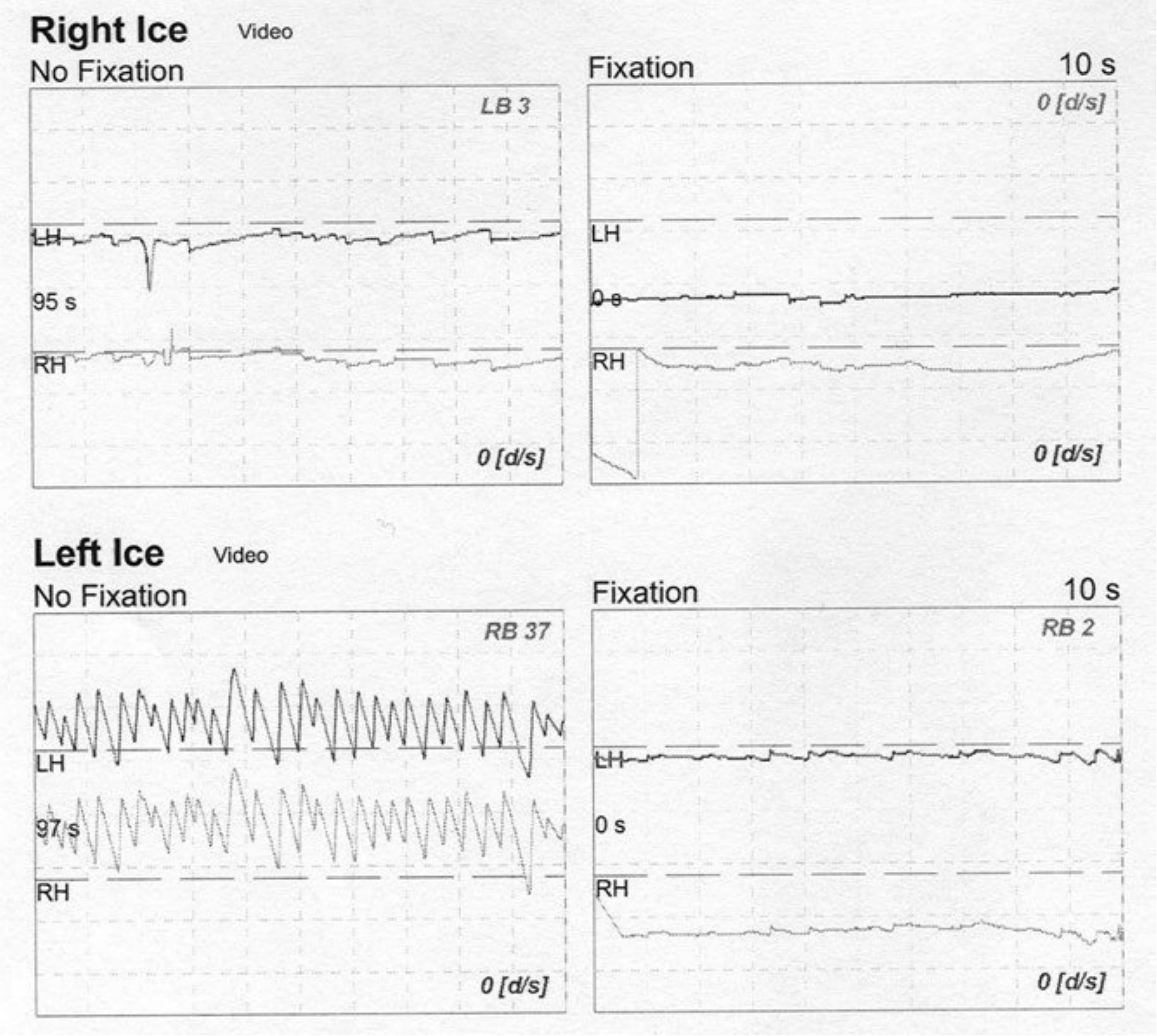

Fig. 36.1 Ice water caloric irrigation results for the right and left ears. This represents the peak response. There is a low (3°/s) response from the right ear and a greater response (37°/s) from the left ear.

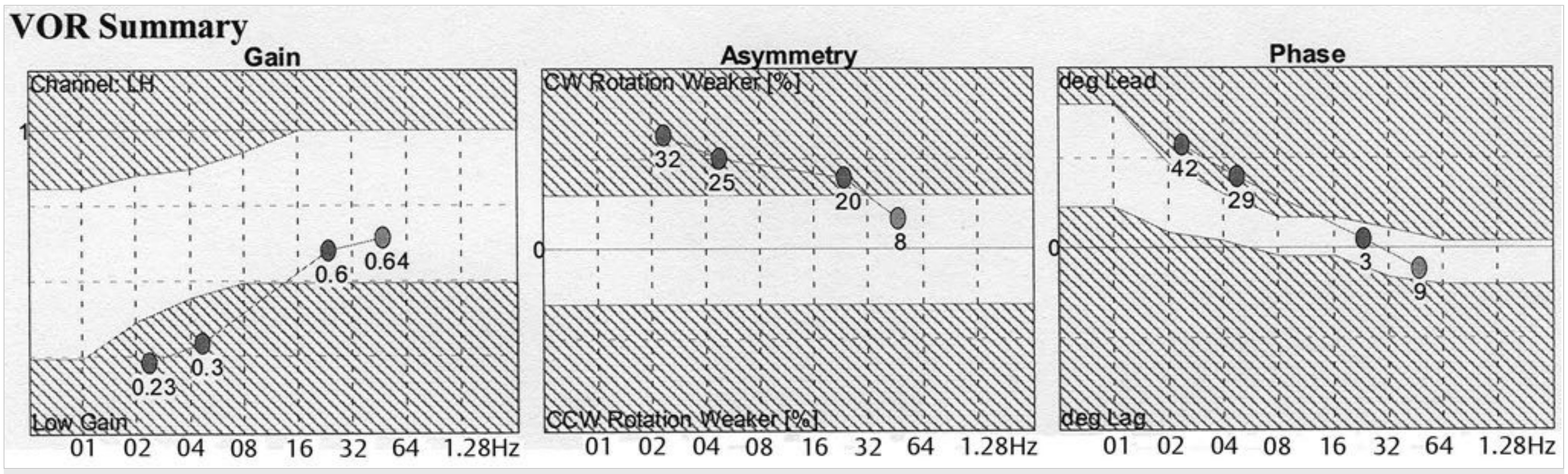

Fig. 36.2 Rotational chair vestibulo-ocular reflex summary. Gain, asymmetry, and phase results are plotted for 0.025, 0.05, 0.25, and 0.5 Hz. There is a low-frequency decrease in gain, and asymmetry with clockwise (rightward) rotation response weaker than counterclockwise (leftward). A phase lead was noted.

counterclockwise rotations (Fig. 36.2). Step velocity time constants (Tc) were reduced. cVEMP responses were obtained and replicated at 95 dB nHL with thresholds of 85 dB nHL and 90 dB nHL in the left and right ears, respectively.

36.3 Questions to the Reader

1. Why did the medical history and clinical exam suggest a diagnosis of bilateral peripheral dysfunction?
2. Did diagnostic testing support the working diagnosis of bilateral peripheral dysfunction?
3. How may unilateral peripheral vestibular disorders present as a bilateral weakness?
4. Was there any information in his history and medical examination that could be explained by unilateral dysfunction?

36.4 Discussion of Questions to the Reader

1. Why did the medical history and clinical exam suggest a diagnosis of bilateral peripheral dysfunction?
   PD complained of his world “jiggling” when he handled power tools, chewed, or moved his head quickly. Bilateral peripheral dysfunction is often accompanied by symptoms of oscillopsia. Oscillopsia is the loss of visual stabilization with head movement. The VOR stabilizes images on the retina; however, when vestibular dysfunction is present the images slip from the retina when the head moves. Oscillopsia makes it difficult to read road signs when riding in the car and makes the environment appear blurry when walking quickly or moving one’s head.

   PD also complained of difficulty walking in the dark. To maintain balance, visual, vestibular, and somatosensory inputs are used. When one of these systems is impaired, such as in the case of vestibular dysfunction, the patient becomes more dependent upon the use of visual and somatosensory cues. If a second of these sensory inputs is compromised, as in the case of walking in the dark, this compounds the impairment. The same is true if somatosensory input becomes incongruent (e.g., walking in sand or on thick rugs). Complaints of having difficulty walking in the dark are common in patients with bilateral vestibular dysfunction. A decrease of greater than seven lines on the Snellen eye chart during DVAT is significant and is consistent with bilateral vestibular dysfunction. A decrease of three or more lines is considered abnormal. A decrease of three to five lines is often seen in unilateral dysfunction, and six or more lines is often seen in bilateral dysfunction. PD had increased sway when standing with eyes closed. When placed on 3-inch foam and asked to close his eyes, he fell with a delayed posture response. With his eyes closed and the foam not allowing appropriate somatosensory input, he had only his vestibular system to rely upon for sensory input. His delayed response implied dysfunction in vestibular input. PD did not realize he was falling until it was too late to catch himself without assistance. The above information in PD’s medical history and the length of time that has passed without improvement supported a diagnosis of bilateral peripheral vestibular dysfunction.
2. Did diagnostic testing support the working diagnosis of bilateral peripheral dysfunction?
   Bithermal caloric responses were bilaterally reduced with a total eye speed of 12°/s. This suggested bilateral peripheral dysfunction. During ice water stimulation, however, there was a marked asymmetry with the right ear being significantly less responsive than the left (R = 3°/s, L = 37°/s) (Fig. 36.1). This suggested asymmetry in the peripheral labyrinthine input with the right side being weaker than the left side.

   RC testing revealed decreased low-frequency VOR gain at 0.025, 0.05 with phase lead. There was an asymmetry with clockwise (rightward) rotations being weaker than counterclockwise (leftward) rotations (Fig. 36.2). Step velocity time constants (Tc) were borderline reduced implying peripheral vestibular dysfunction. The decreased low-frequency VOR gain with phase lead can be seen in either bilateral peripheral dysfunction or uncompensated unilateral dysfunction. The asymmetry in the VOR response revealed responses were weaker with the clockwise (rightward) rotations as compared with the counterclockwise rotations (leftward). This finding suggested asymmetry in the peripheral labyrinthine input with the right side being weaker than the left side.

cVEMP responses were obtained and replicated at 95 dB nHL. A threshold of 85 dB nHL was obtained on the left and a threshold of 90 dB nHL was obtained on the right. This suggests normal threshold levels bilaterally.

Overall, the asymmetric results in the ice water test and the asymmetry in VOR responses during RC testing suggested that the right ear was weaker than the left ear.

3. How may unilateral peripheral vestibular disorders present themselves as a bilateral weakness?
During the acute phase of a unilateral peripheral insult there is a sudden decrease in activity from one vestibular nucleus. The brain recognizes this pattern of asymmetric input and responds as if the patient is indeed spinning in the direction of the intact side. Proprioceptive cues send conflicting information to the brain that the patient is indeed not moving. The brain responds by decreasing the nuclear activity of the vestibular nuclei bilaterally with the most inhibition taking place within the noninvolved vestibular nucleus. This dampening, or central inhibition of the functioning labyrinth, is thought to be a cerebellar function and is the brain's first protective reaction with the intent to reestablish symmetry within the system. This decreases symptoms and the mismatch of information being transmitted to the brain. Soon after this central inhibition, a regeneration of activity in the nucleus of the affected ear begins. This is the beginning of the compensation process. When the affected nucleus has regenerated its resting activity, static compensation has been achieved. No symptoms are perceived as long as there is no movement of the head. This process is accomplished automatically. PD experienced symptoms when he moved his head, however, and needed exercises prescribed by a vestibular rehabilitative professional in order to complete his dynamic compensation.
4. Was there any information in his history and medical examination that could be explained by unilateral dysfunction?
The blurring vision when using the handheld power tools may be stimulating a vibration-induced nystagmus. Vibration is sometimes applied to the skull or neck during DVAT testing in an effort to uncover a unilateral vestibular deficit. PD revealed a head impulse sign during HIT in the right horizontal plane. This suggested right horizontal canal dysfunction.

36.5 Diagnosis and Recommended Treatment

PD's diagnosis was right unilateral vestibular dysfunction. Vestibular rehabilitation for uncompensated unilateral dysfunction was planned.

36.6 Outcome

PD is currently enrolled in a vestibular rehabilitation/retraining program in his home state. He is showing improvement in dynamic compensation.

36.7 Key Points

1. A well-taken history, medical exam, and diagnostic battery are all essential to the diagnosis and management of the balance-disordered patient.
2. Compensation occurs in two phases. In the first phase, there is suppression of the static asymmetry. In the second phase, neural activity during head movement occurs.
3. Unilateral vestibular dysfunction can create oscillopsia with rapid head movement.

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37 Differential Diagnosis of Central versus Peripheral Vertigo

Hillary Snapp

A 51-year-old female presents to the clinic with a 5-month history of dizziness and imbalance.

37.1 Clinical History and Description

EM is a 51-year-old female who was referred for vestibular evaluation. EM presented with a complicated medical history including recent diagnosis of celiac disease, severe anemia requiring blood transfusion, shingles, and bradycardia (abnormally low resting heart rate) resulting in hospitalization. EM also had a significant history of alcohol and drug abuse 17 years earlier resulting in seizure disorder and bipolar disorder regulated with lithium. EM reported that loud sounds can trigger her seizures. EM's medical history was also positive for migraines regulated with sumatriptan. EM was involved in a car accident 20 years ago resulting in loss of consciousness and subdural hematoma. EM reports that 1 month following the car accident she was told she had acute labyrinthitis and was prescribed meclizine which EM took daily for 9 months. Antivert is a vestibular suppressant that is typically used to alleviate vertigo during the acute phase. It is not intended for long-term use because it can result in chronically uncompensated vestibulopathy.

Vestibular complaints at the time of presentation included 5 months of nausea and vomiting, feeling "drunk" upon rising or walking, needing to hold on to the wall while walking, and falls. Dizziness was essentially constant, but more severe in the morning. EM reported she was performing daily positional exercises prescribed by an outside facility that did not improve symptoms. The exercises were described as maneuvers such as rapid head turns right and left and rolling from side to side while lying in a supine position. EM's dizziness could be triggered by loud sounds (Tullio phenomenon) as well as activities involving exertion. EM reported a feeling of falling or "being pulled to earth," but denied true vertigo, hearing loss, tinnitus, otalgia, aural fullness, diabetes, motion sickness, or previous ear surgery.

EM was referred to audiology for audiological and vestibular examination and her neurotologist ordered magnetic resonance imaging (MRI) of the internal auditory canals and brain.

37.2 Audiological Testing

The audiometric examination (□ Fig. 37.1) revealed a mild bilateral conductive hearing loss at 250 Hz with an air–bone gap of 30 dB HL in the right ear and 20 dB HL in the left ear. For the right ear the audiometric configuration rose to a slight hearing loss at 500 and 4000 Hz as well as hearing being within normal limits at 1000 to 2000 Hz and 8000 Hz. Results for the left ear in normal pure-tone thresholds at 250, 1000, and 4000 Hz and a slight hearing loss at 500, 2000, and 8000 Hz. Speech recognition thresholds (SRTs) and word recognition scores (WRSs) were within normal limits bilaterally. Immittance audiometry revealed bilaterally normal tympanograms and normal ipsilateral and contralateral acoustic reflex thresholds (ARTs).

37.3 Videonystagmography Examination

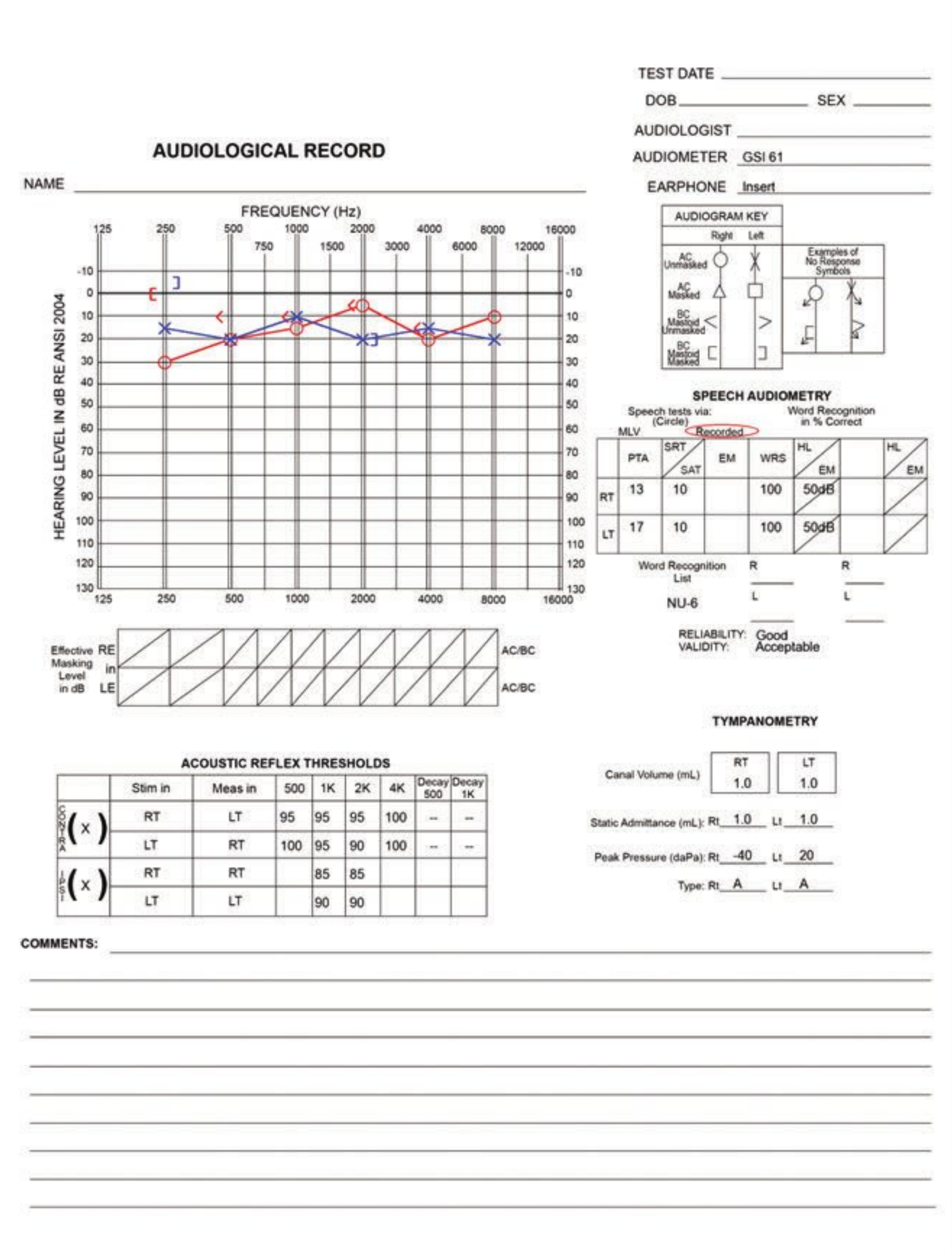
The videonystagmography (VNG) examination revealed no spontaneous or gaze-evoked nystagmus and unremarkable oculomotor and positional testing. Robust saccadic eye movement was observed throughout testing. High-frequency headshake resulted in significant downbeat nystagmus post-rotation. The Dix-Hallpike maneuver was unremarkable; however, EM became very disoriented upon sitting from the head hanging right position and had to be stabilized. Bithermal caloric testing using water stimulus resulted in an asymmetry calculation of 23% with a right reduced vestibular response compared to left.

37.4 Questions to the Reader

1. Based on the reported case history, what are the diagnostic considerations? What does the case history lead the audiologist to consider for differential diagnosis?
2. Are there any significant findings from the audiological examination to assist the clinician in differential diagnosis?
3. Do the results from the audiological examination provide any valuable information to the vestibular assessment?
4. How might EM's medical history impact the vestibular testing?
5. What are the significant findings seen during the vestibular testing? Are there any key points in EM's case history that relate to the findings?
6. What would a clinician recommend based on these results from the VNG examination?

37.5 Discussion of Questions to the Reader

1. Based on the reported case history, what are the diagnostic considerations? What does the case history lead the audiologist to consider for differential diagnosis?
   EM's case history does not provide the audiologist with a clear clinical pattern. There are many symptoms within EM's case history to consider. The report of constant unsteadiness and absence of true vertigo may suggest a central nervous system (CNS) disorder, which is consistent with EM's history of migraines, seizures, long-term drug and alcohol abuse, and use of lithium. EM, however, reports peripheral vestibular indicators such as previous diagnosis of acute labyrinthitis



AUDIOLOGICAL RECORD

NAME

TEST DATE

DOB SEX

AUDIOLOGIST

AUDIOMETER GSI 61

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	13	10		100	50dB		
LT	17	10		100	50dB		

Word Recognition List NU-6 R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.0	1.0

Static Admittance (mL): Rt 1.0 Lt 1.0

Peak Pressure (daPa): Rt -40 Lt 20

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	95	95	95	100	--	--
	LT	RT	100	95	90	100	--	--
IPSI (x)	RT	RT		85	85			
	LT	LT		90	90			

COMMENTS:

Fig. 37.1 Audiological examination demonstrating normal hearing and a bilateral air–bone gap at 250 Hz.

and dizziness that is evoked by loud sounds (Tullio phenomenon) or changes in intrathoracic pressure. This is commonly described as dizziness evoked by sneezing, lifting heaving objects, exercising, or exertion with constipation. Differential diagnosis should include pharmacologically induced dizziness related to her long-term use of lithium, vestibular migraine, and peripheral disorders associated with the Tullio phenomenon. The Tullio phenomenon, combined with changes in intrathoracic pressure, suggests a third window effect such as a fistula or dehiscence of the vestibule. This refers to the change in resistance of the auditory system due to an abnormal opening in the bony structure, thereby resulting in changes to hearing and balance sensitivity.

2. Are there any significant findings from the audiological examination to assist the clinician in differential diagnosis?
   Review of the audiogram reveals a mild low-frequency conductive hearing loss in the right ear, and although hearing sensitivity remains within normal limits on the left ear, a significant air–bone gap is observed at 250 Hz bilaterally. Tympanograms, however, are normal bilaterally.
3. Do the results from the audiological examination provide any valuable information to the vestibular assessment?
   In the absence of any other indications of middle ear pathology (i.e., normal tympanograms and normal ARTs) the bilateral air–bone gap at 250 Hz is consistent with a third window effect. A pathological third window effect reduces the impedance of the cochlear fluids thereby improving the cochlear response to bone conduction and resulting in supranormal bone conduction thresholds.
4. How might EM's medical history impact the vestibular testing?
   Lithium is a medication that acts on the CNS. As such, EM's long-term use of multiple medications including lithium for more than 15 years can impact the function of the CNS. Large dosages and/or long-term use of lithium can cause cerebellar toxicity and result in central nystagmus such as periodic alternating nystagmus or downbeat nystagmus. Additionally, EM had a history of acute labyrinthitis treated with Antivert for approximately 9 months. As stated previously, Antivert is a vestibular suppressant that is typically used to alleviate vertigo during the acute phase. Following an acute

vertiginous episode, most peripheral vestibular disorders will resolve naturally in about 6 to 10 weeks as the brain works to restore balance to asymmetric neural firings of the balance organs. This process is termed vestibular compensation. Long-term use of vestibular suppressants prevents the natural central compensation process from taking place and may result in chronic dizziness and/or imbalance. This, however, is not suspected to be related to EM's current complaints of dizziness because her treatment with Antivert was 20 years prior to the onset of her current symptoms. It is important for the clinician to be aware of the potential interaction of these comorbidities (long-term use of medication and acute labyrinthitis) on the test results and to discern underlying preexisting conditions from the findings related to the current patient complaints as best as possible.

5. What are the significant findings seen during the vestibular testing? Are there any key points in EM's case history that relate to the findings?
   The known pharmacological influence of the lithium is likely responsible for the ongoing saccadic eye movement observed during testing and therefore is not pathological in nature. Abnormal findings on tests of oculomotor function generally suggest pathologies of central origin. Saccades are a central phenomenon produced and controlled primarily by the brainstem and cerebellum. Because of this, drugs that act on the CNS can result in involuntary oscillations or movements of the eyes that are not truly pathological in nature. This is why it is important to thoroughly review all long-term and short-term medications reported by the patient during the case history. In this case, the clinician can determine that oculomotor function is otherwise intact. EM also presents with downbeating nystagmus, which is typically considered a central indicator; however, this may also be a result of lithium toxicity
   EM's bithermal caloric responses were borderline significant for unilateral weakness at 23%asymmetry. A unilateral vestibular weakness is defined as an asymmetric caloric response of>25%between ears. The clinician should consider how this finding may be a result of EM's previous reported episode of acute labyrinthitis and may be unrelated to the current patient reports. Additionally, EM had been completing rehabilitative positioning exercises at home, which failed to improve her symptoms. In the case of a unilateral horizontal canal lesion, such exercises would be expected to assist the compensation process and improve the symptoms of dizziness. It may be that the centrally acting medications EM is taking (i.e., lithium) are limiting this compensation process, or the horizontal semicircular canal may not be involved.
6. What would a clinician recommend based on these results from the VNG examination?
   EM's dizziness is primarily triggered by loud sounds or changes in intrathoracic pressure, which were not assessed at this visit. As such, Tullio and fistula testing were recommended. Tullio testing consists of presenting the patient with a loud acoustic stimulus (usually a pure-tone signal using an insert earphone), and recording the eyes for onset of nystagmus linked to the acoustic stimulus. Fistula testing consists of presenting positive and negative pressure to the ear canal while recording the eyes for onset of nystagmus after pressurization.
   Cervical vestibular-evoked myogenic potential (cVEMP) testing was recommended because of the suspicion of a third window effect (i.e., presence of air–bone gaps at 250 Hz with normal tympanometry and reported Tullio phenomenon). Additionally, EM reports a sensation of falling/being pulled to the earth, which may suggest otolith involvement because the otoliths are responsible for movement in the linear plane, whereas the semicircular canals are responsible for angular movements.

37.6 Vestibular Testing

The cVEMP is a sonomotor response recorded from the sternocleidomastoid (SCM) muscle in the neck in response to a high-intensity acoustic stimulus (100 dB nHL) delivered via an insert earphone. Recording electrodes are placed at each SCM. The reference electrode is placed on the ipsilateral SCM, and the active electrode is placed on the contralateral SCM with a ground electrode placed at the forehead. The electrophysiologically recorded response (□ Fig. 37.2) consists of a primary peak at a latency of 13 ms (P1) followed by a negative peak at 23 ms (N1). This response represents the vestibulocollic reflex and provides information regarding function of the saccule and inferior branch of the vestibular nerve.

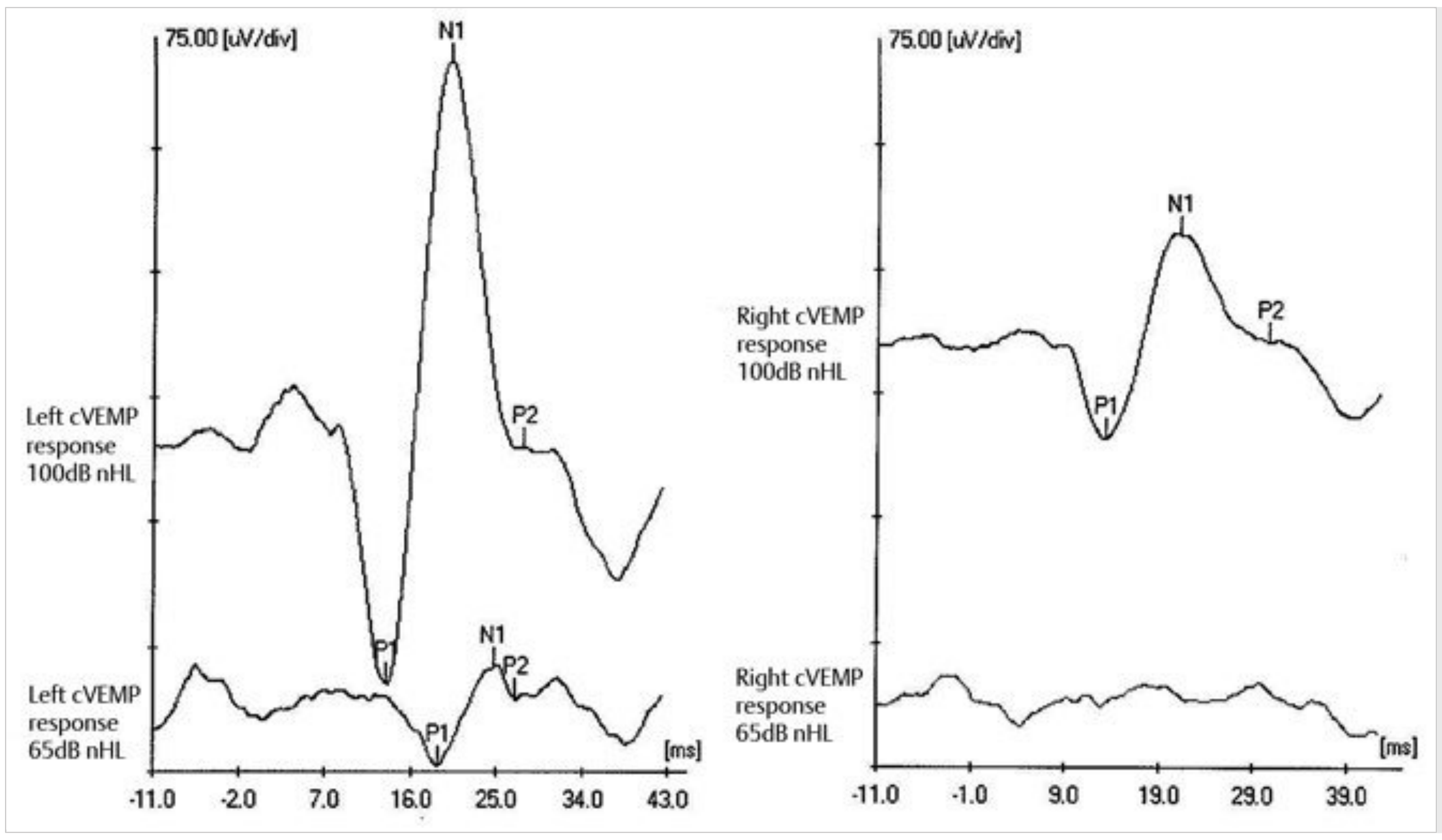


Fig. 37.2 Cervical vestibular-evoked myogenic potential (cVEMP) test results demonstrating a 51% asymmetry ratio of left increased amplitude compared with right and positive threshold response in the left ear at 65 dB nHL. Normal threshold responses are observed at ≥ 70 dB nHL. You can see by the tracings that the right response is eliminated below 70 dB nHL whereas the left response remains intact. Note that this cVEMP is recorded "upside down" or inverted where the negative trough represents the positive deflections (P_1 & P_2) and the positive trough represents the negative deflection (N_1). For this figure, the y-axis represents amplitude in microvolts (μV), and the x-axis represents time in milliseconds (ms).

EM's cVEMP testing resulted in a significant asymmetry ratio of 51% with left amplitude being greater than right amplitude. A reduced amplitude threshold response was also observed in the left ear. Normal threshold responses are observed at 70 to 90 dB nHL, and asymmetry should be less than 36% to 45% The Tullio test was not performed because EM reported extreme vertigo during left cVEMP testing, which is consistent with the Tullio phenomenon. To visualize if nystagmus was present during cVEMP testing, Frenzel goggles were employed and a robust rotational nystagmus was observed for the duration of the stimulus, even at threshold during the cVEMP test. The patient became extremely disoriented and had to discontinue testing. As a result of this, fistula testing was not completed.

37.7 Additional Questions to the Reader

1. EM reported an onset of seizures with loud sounds. Should this have been considered prior to the recommendation of cVEMP and Tullio testing?
2. Where do the results from these examinations lead the clinician?

37.8 Discussion of Additional Questions to the Reader

1. EM reported an onset of seizures with loud sounds. Should this have been considered prior to the recommendation of cVEMP and Tullio testing?
   It is important to note in the patient's electronic medical record (EMR) any complications such as those reported in this case report, and it is also important to discuss these with the patient and the referring physician prior to testing. In this case, it became clear through several discussions with EM that she likely had been having severe vertiginous attacks following the introduction of loud sounds that were previously misdiagnosed. EM was advised of the stimulus used during cVEMP and Tullio testing and agreed to the examinations, which were conducted in a medical facility staffed by nurses and physicians who could respond in the event of an emergency.
2. Where do the results from these examinations lead the clinician?
   Despite the patient case history, EM's symptoms and test results do not suggest CNS involvement. Improved bone conduction thresholds (i.e., air–bone gap) in the presence of normal tympanograms and ARTs, the presence of the Tullio phenomenon, and dizziness triggered by intrathoracic pressure are consistent with third window effect. Differential diagnosis includes assessing if EM's symptoms are related to a perilymphatic fistula and/or dehiscence of the superior semicircular canal (DSSC). Perilymphatic fistula most commonly occurs at the oval or round window and is commonly associated with hearing loss and tinnitus, both of which were denied by EM. In this case, the presence of the headshake-evoked downbeat nystagmus was not suspected to be central in nature and is consistent with superior semicircular canal (also known as the anterior semicircular canal) involvement.
   Further, cVEMP is a test of the saccule, and inferior branch of the vestibular nerve. The superior semicircular canal is innervated by the inferior branch of the vestibular nerve. Results of a positive cVEMP test with a reduced threshold response is consistent with a third window or DSSC and is unlikely related to a perilymphatic fistula. The reduced threshold is observed due to an increase in sensitivity to sound resulting from the change in resistance of the bony structure of the auditory-vestibular system. Based on the reported findings, a computed tomographic (CT) scan was ordered to assess the bony structure of the superior semicircular canals and to confirm or deny the presence of a DSSC.

37.9 Diagnosis and Recommended Treatment

Comprehensive audiological and vestibular examination revealed a pattern consistent with left DSSC. The presence of downbeating nystagmus post-headshake was suggestive of superior semicircular canal involvement. The Tullio phenomenon was observed during sound stimulation to the left ear, evidenced by observable nystagmus and reported subjective dizziness. An increased cVEMP amplitude was observed in the left ear compared with the right ear, with a reduced threshold indicating third window syndrome of the superior semicircular canal. The MRI scan was negative. Surprisingly, the CT scan of the temporal bones revealed bilateral DSSC. This finding is consistent with the audiological examination, which revealed bilateral suprathreshold bone conduction responses as evidenced by bilateral air–bone gaps at 250 Hz, and normal tympanograms and ARTs.

In summary, EM presents with symptoms of third window syndrome, particularly a left DSSC. EM does have some dehiscence bilaterally; however, her left vestibular system appears to be the symptomatic ear. As a result of the audiological and vestibular examination findings and radiographic tests, EM was scheduled for left superior semicircular canal plugging via a middle fossa craniotomy. This surgical approach consists of opening the skull above the ear to identify the semicircular canal dehiscence. The dehiscence can either be plugged or resurfaced. Plugging is often considered to be more effective at repairing the dehiscence, but it also carries a higher risk of sensorineural hearing loss. Many individuals may choose to manage the DSSC through lifestyle modifications to reduce evoking the symptoms rather than pursuing surgery.

37.10 Outcome

At 1 week postsurgery, EM reported significant improvement in her symptoms, but she still reported occasional vertigo. On postsurgical vestibular evaluation, EM presented with mild right beat nystagmus post-headshake and downbeating nystagmus in the head-hanging position (test of anterior/superior semicircular canal). The reduced left threshold response was no longer present during cVEMP testing, suggesting results are now within normal limits. Bithermal caloric irrigations indicate no change in horizontal semicircular canal function. EM's neurotologist diagnosed EM with postural nystagmus due to a superior semicircular canal defect, which is an expected outcome following the surgical plugging of the superior semicircular canal. EM's associated comorbidities (CNS-acting medications) delayed vestibular compensation, resulting in additional vestibular rehabilitation therapy. At 6 months postoperative there was no evidence of downbeating nystagmus on headshake or head-hanging positional testing. Additionally, EM was fully resolved of all symptoms, denied falls, and was able to walk unassisted.

This case report represents the importance of careful evaluation and management of patients who present with potential comorbidities. Not only does this complicate the clinician's diagnosis but it often impacts a patient's ability to compensate following treatment for peripheral lesions.

37.11 Key Points

1. Dehiscence of the bone covering the semicircular canals can result in vertigo and/or changes in hearing sensitivity.
2. Centrally acting medications can interfere with test results and mislead the clinician. Understanding how these medications may impact the peripheral and central vestibular systems is important for accurate interpretation of test findings.
3. Downbeat nystagmus is most commonly associated with disorders of the central vestibular system; however, it is also seen in pathology of the superior canal.
4. Diagnosis of vestibular disorders is often complex and rarely straightforward. An in-depth knowledge of the anatomy and physiology of the vestibular system as well as the associated oculomotor system and central mechanisms is critical to the successful differential diagnosis of patients complaining of dizziness.
5. A comprehensive case history is a fundamental component of the diagnostic process and can expose any comorbidity that may affect performance or interpretation of test findings.

Suggested Reading

Jacobson GP, McCaslin DL, Piker EG, Gruenwald J, Grantham SL, Tegel L. Patterns of abnormality in cVEMP, oVEMP, and caloric tests may provide topological information about vestibular impairment. J Am Acad Audiol 2011; 22: 601–611

Jongkees LB, Maas JP, Philipszoon AJ. Clinical nystagmography: a detailed study of electro-nystagmography in 341 patients with vertigo. Pract Otorhinolaryngol (Basel) 1962; 24: 65–93

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Williams DP, Troost BT, Rogers J. Lithium-induced downbeat nystagmus. Arch Neurol 1988; 45: 1022–1023

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38 Identification of Superior Semicircular Canal Dehiscence

Linsey Scheibler, Michael J. Cevette, and David Barrs

A 39-year-old female was referred for vestibular evaluation due to unsteadiness and inability to visually fixate, especially with quick head turns.

38.1 Clinical History and Description

PG was a 39-year-old female who presented to an audiology clinic due to increasing unsteadiness and loss of balance with quick head movements over the past 5 years. She also noted that her ability to visually fixate was impaired (i.e., oscillopsia) or "did not keep up" especially when she was running or walking on uneven surfaces. As a result, she experienced difficulty monitoring her students' computer screens during her work as an instructor in a media center. Otologic reports included aural fullness in the left ear and a loud heartbeat sound in her head, but not isolated to either ear. PG denied any true vertigo, tinnitus, otalgia, or history of noise exposure. Her current medications included duloxetine and alprazolam. Previous records from an outside facility indicated a normal audiological examination and unremarkable findings for electroencephalography (EEG), computed tomographic (CT) scan, magnetic resonance imaging (MRI), and magnetic resonance angiography (MRA). As part of her care at the audiology clinic, PG was referred for a comprehensive audiological and vestibular examination including videonystagmography (VNG), cervical vestibular-evoked myogenic potentials (cVEMPs), and computerized dynamic posturography (CDP).

38.2 Audiological Testing

The results of the comprehensive audiological examination revealed air conduction thresholds within normal limits bilaterally but with notable air–bone gaps from 250 to 2000 Hz in the left ear and at 250 Hz in the right ear. Left masked bone thresholds were present at the lowest limit of the audiometer, − 10 dB HL, resulting in the air–bone gaps. Speech recognition thresholds (SRTs) were evaluated using recorded spondees and were consistent with air thresholds bilaterally. Word recognition scores (WRSs) were completed using recorded isophonemic word lists and revealed 100% word recognition bilaterally. Tympanometry results were consistent with type A tympanograms bilaterally, showing both pressure and static compliance values within normal limits (between − 100 and + 100 daPa for pressure and between 0.3 and 2.0 mL for compliance). Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present at normal hearing levels bilaterally (Fig. 38.1).

CDP testing was completed using the NeuroCom Equitest Computerized Dynamic Posturography equipment (NeuroCom, Clackamas, OR). Testing included the motor control test (MCT), adaptation test, and sensory organization test (SOT). PG's composite SOT score was 74, with scores greater than 70 considered within normal limits based on available age-matched normative data in the NeuroCom software. The MCT and adaptation tests also yielded latencies within normal limits. VNG testing included oculomotor assessment (spontaneous, gaze, smooth pursuit, and saccadic tracking), the headshake test, positioning assessment (Dix-Hallpike and lateral head rolls), static positional testing, and bithermal water caloric irrigations. All findings were normal, including caloric stimulation.

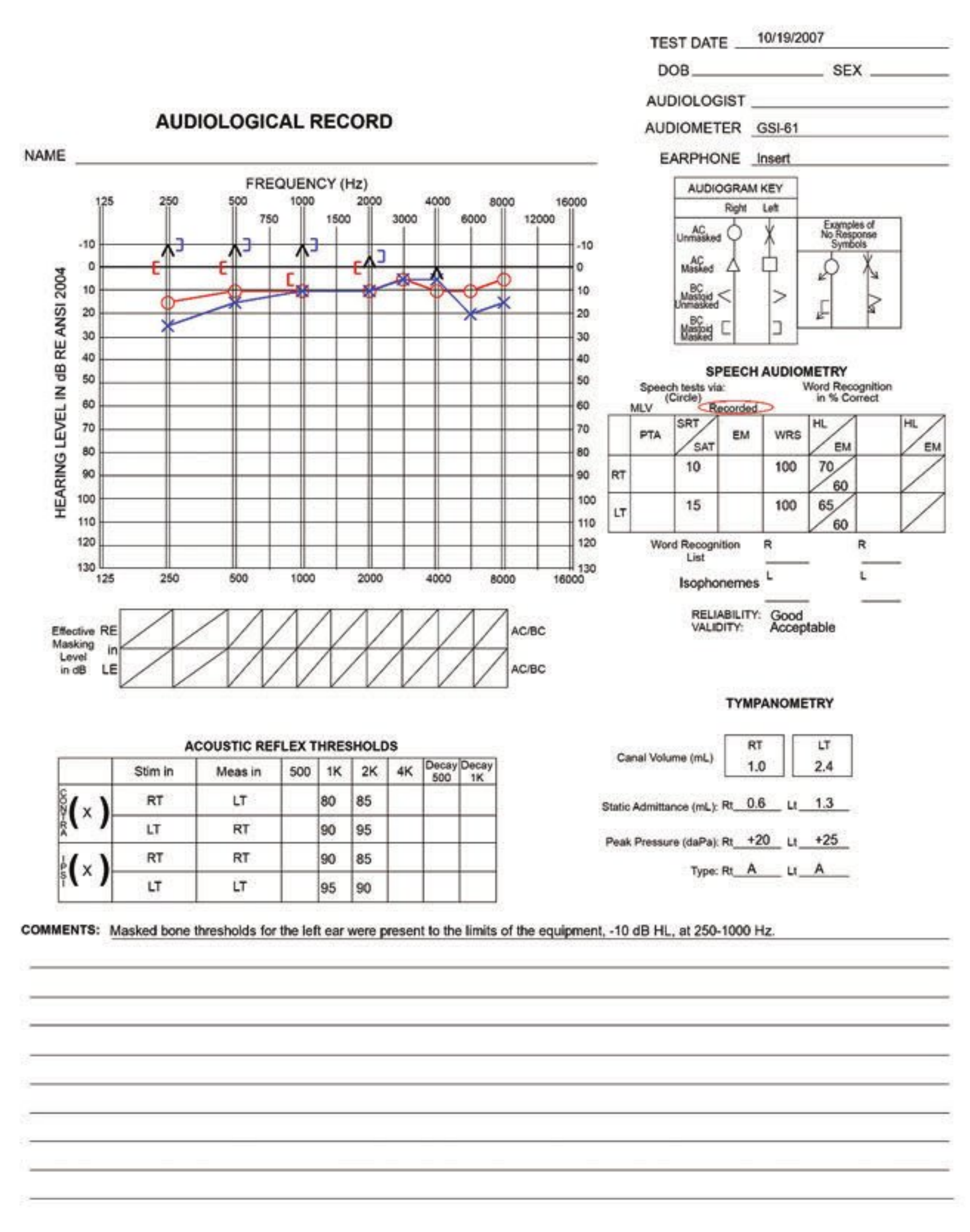
The cVEMP was measured at 98 dB nHL using a 500 Hz tone burst. The active electrode was placed in the center of the forehead with the larger reference electrode on the upper third of the sternocleidomastoid muscle. A ground electrode was placed above the eyebrow, approximately one and a half inches from the active electrode. The muscle contraction was initiated by PG lifting her head off the head rest from a 30° supine position with her head turned away from the test ear. P1 and N1 latencies were within normal limits (< 20 ms and < 30 ms, respectively, based on clinic normative data) as was the interaural amplitude difference (measured at 22% greater in the left ear with > 47% considered a significant asymmetry based on clinic normative data). The VEMP thresholds were 75 dB nHL and 60 dB nHL for the right and left ears, respectively, resulting in a significant 15 dB interaural threshold difference (> 10 dB interaural difference is considered abnormal based on clinic normative data).

38.3 Questions for the Reader

1. Which audiological findings support a diagnosis of superior semicircular canal dehiscence (SSCD)? Does the patient's case history also support this diagnosis and why?
2. Explain the proposed mechanism causing the air–bone gaps and lower cVEMP thresholds in patients with SSCD.
3. What additional testing might be considered for this patient and why?

38.4 Discussion of Questions to the Reader

1. Which audiological findings support a diagnosis of SSCD? Does the patient's case history also support this diagnosis and why?
   The notable better-than-normal bone conduction thresholds for the left ear in the presence of normal tympanograms and ARTs correlates highly with SSCD. Additionally, the 15 dB lower cVEMP threshold when measured on the left side compared with the right side further supports a diagnosis of SSCD in the left ear. PG's case history including chronic imbalance, aural fullness in the left ear, perception of audible heartbeat, and oscillopsia represents a constellation of symptoms associated with SSCD. CT scans are often considered the gold standard for identification of SSCD and have even been found to overestimate its prevalence compared with pathological studies. Therefore, the presence of the normal CT scan would exclude a diagnosis of SSCD in this case.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HEARING LEVEL IN dB RE ANSI 2004
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AUDIOGRAM KEY
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AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
Effective Masking Level in dB
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in
LE
AC/BC
AC/BC

AUDIOLOGICAL RECORD

TEST DATE 10/19/2007

DOB

SEX

AUDIOLOGIST

AUDIOMETER GSI-61

EARPHONE Insert

NAME

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		10		100	70 / 60		
LT		15		100	65 / 60		

Word Recognition List R L R L

Isophonemes

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.0	2.4

Static Admittance (mL): Rt 0.6 Lt 1.3

Peak Pressure (daPa): Rt +20 Lt +25

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT		80	85			
	LT	RT		90	95			
IPSI (X)	RT	RT		90	85			
	LT	LT		95	90			

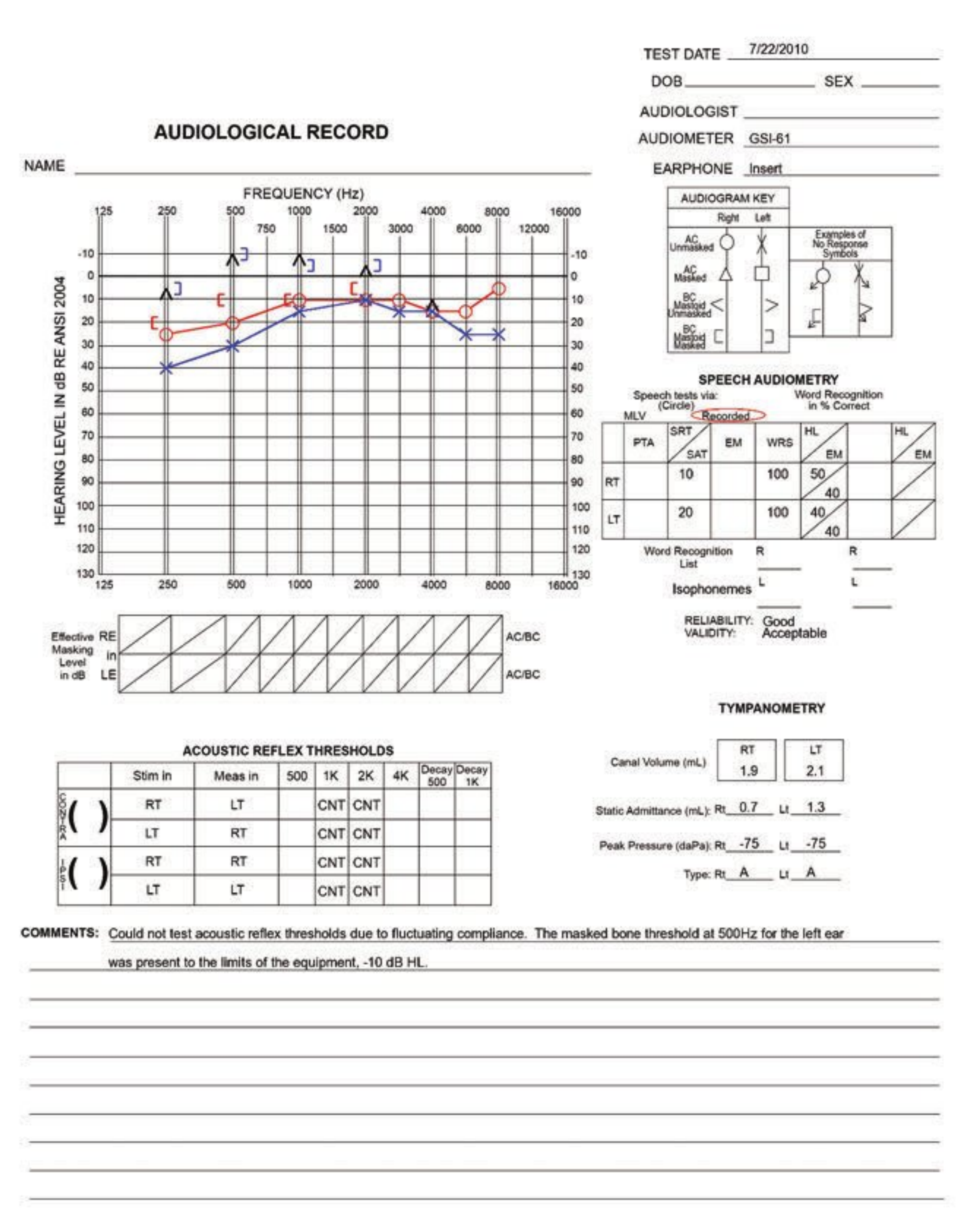
COMMENTS: Masked bone thresholds for the left ear were present to the limits of the equipment, -10 dB HL, at 250-1000 Hz.

Fig. 38.1 Initial audiological examination reporting normal air conduction thresholds with large air–bone gaps in the left ear (35 dB HL at 250 Hz, 25 dB HL at 500 Hz, 20 dB HL at 1000 Hz, and 15 dB HL at 2000 Hz) and normal tympanograms and acoustic reflex thresholds to contralateral and ipsilateral stimulation.

2. Explain the proposed mechanism causing the air–bone gaps and lower cVEMP thresholds in patients with SSCD.
   The “third window,” or opening in the superior semicircular canal (SSC) in addition to the oval and round windows of the cochlea allows for sound-induced motion of the fluid within the SSC. Because the inner ear fluid is displaced from the cochlea to the semicircular canal during an air-conducted signal there is a decrease in sound pressure that is present to activate the cochlea and therefore result in poorer air conduction thresholds. The dehiscent semicircular canal also allows for a lower resistance to bone-conducted sounds and therefore may cause better-than-expected bone conduction thresholds. The decreased cVEMP thresholds to air-conducted stimuli are also a result of the third window allowing fluid motion within the vestibular system compared with an intact SSC.
3. What additional testing might be considered for this patient and why?
   The clinician might consider evaluating PG using an ocular VEMP (oVEMP). oVEMP amplitudes may provide greater sensitivity and specificity for identification of SSCD compared with cVEMP threshold results. Also, oVEMP amplitudes are quicker to measure than cVEMP thresholds. When compared to measuring the cVEMP, oVEMP only requires that the patient maintain an upward gaze instead of holding a sternocleidomastoid muscle contraction. The latter requirement can be uncomfortable for some patients. In addition, this contraction can be difficult to reliably repeat between trials.

38.5 Additional Testing

Due to the conflicting findings between the normal CT, the case history, and audiological findings the otologist obtained the original CT scan images for reanalysis by a neuroradiologist. The second interpretation of the images by the neuroradiologist suggested thinning of the superior canal bilaterally, but the neuroradiologist felt a definitive diagnosis could not be made due to inadequate scan resolution.


FREQUENCY (Hz)
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HEARING LEVEL IN dB RE ANSI 2004
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130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols

AUDIOLOGICAL RECORD

NAME

TEST DATE 7/22/2010

DOB

SEX

AUDIOLOGIST

AUDIOMETER GSI-61

EARPHONE Insert

Effective Masking Level in dB RE in LE AC/BC AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		10		100	50 / 40		
LT		20		100	40 / 40		

Word Recognition List R R

Isophonemes L L

RELIABILITY: Good

VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT		CNT	CNT			
	LT	RT		CNT	CNT			
IPSI ( )	RT	RT		CNT	CNT			
	LT	LT		CNT	CNT			

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.9	2.1

Static Admittance (mL): Rt 0.7 Lt 1.3

Peak Pressure (daPa): Rt -75 Lt -75

Type: Rt A Lt A

COMMENTS: Could not test acoustic reflex thresholds due to fluctuating compliance. The masked bone threshold at 500Hz for the left ear was present to the limits of the equipment, -10 dB HL.

Fig. 38.2 Audiological examination from approximately 3 years later showing a mild low-frequency conductive hearing loss in the left ear and an increase in the maximal air–bone gap in the left ear (40 dB HL at 500 Hz compared with only 25 dB HL at 500 Hz 3 years earlier) and normal tympanograms and acoustic reflex thresholds to contralateral and ipsilateral stimulation.

Despite a recommendation for a repeat CT scan, the patient did not return for reexamination until 3 years later. At that time PG reported increased oscillopsia that prevented her from driving or exercising. She also experienced the Tullio phenomenon (i.e., dizziness in the presence of loud sounds) with sound in the left ear as well as increased aural fullness in the left ear. The audiogram reported an increase in the air–bone gaps in the left ear compared with the audiogram from 3 years prior and no significant air–bone gaps in the right ear (none > 10 dB). Left-sided air–bone gaps had increased to 35 dB HL at 250 Hz, 40 dB HL at 500 Hz, 20 dB HL at 1000 Hz, and 15 dB HL at 2000 Hz (Fig. 38.2). PG underwent a repeat CT scan, which revealed a clear dehiscence on the left side (Fig. 38.3). Vestibular testing was not completed because the otologist felt that a reliable diagnosis could be made using the case history, audiogram, repeat CT scan findings, and previous cVEMP results.

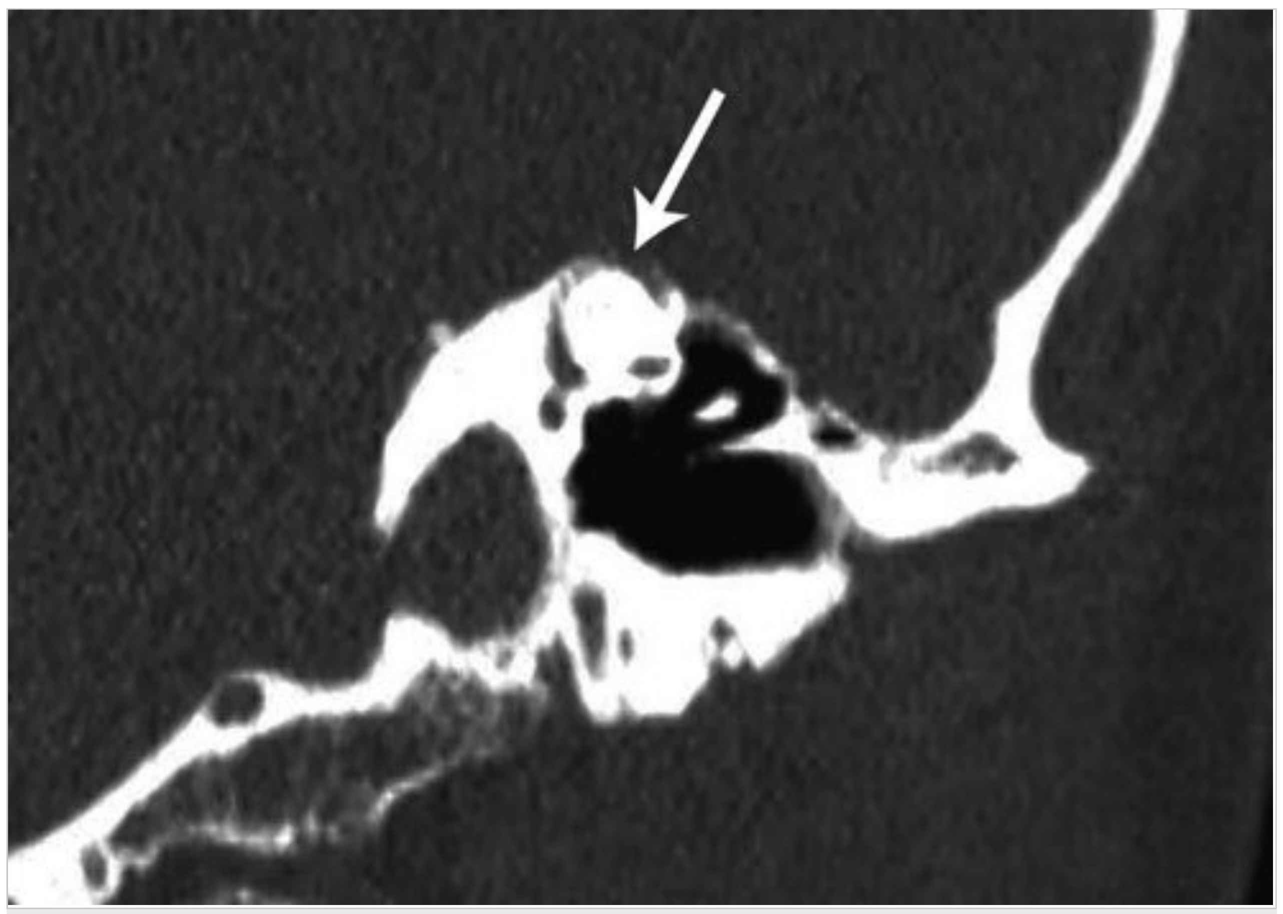

Fig. 38.3 Reconstructed computed tomographic (CT) scan of the left temporal bone perpendicular to the superior semicircular canal showing loss of bone overlying the canal that is consistent with semicircular canal dehiscence. The arrow points to a loss of bone (bone is white on CT scans) overlying the lumen of the superior semicircular canal. Lumen is dark on CT scans and would be completely covered by the white-appearing bone in a scan showing an intact semicircular canal.

38.6 Additional Question to the Reader

1. What evidence suggests that the size of the dehiscence had increased over the last 3 years?

38.7 Discussion of Additional Question to the Reader

1. What evidence suggests that the size of the dehiscence had increased over the last 3 years?
   Although the initial CT scan was deemed unreliable and the second CT scan did not measure the size of the dehiscence, the fact that the air–bone gaps increased would suggest that the size of the dehiscence increased. Although one might suspect that the case history would also suggest an increase in size due to the worsening symptoms, some studies have reported a lack of a correlation between the length of the dehiscence and pure-tone average (PTA), bone conduction thresholds, cVEMP thresholds, and other presenting signs and symptoms. The maximal air–bone gap was the only factor positively correlated with the size of the dehiscence.

38.8 Diagnosis and Recommended Treatment

Based on the diagnosis of left-sided SSCD and the significant difficulty PG was experiencing in her daily living, it was recommended that PG consider surgical intervention. The surgical procedure to repair the SSCD involves either a middle fossa or a transmastoid approach to the dehiscence. This abnormal opening in the semicircular canal is then packed with soft tissue, bone dust, or similar substance and often further protected with a plate of bone or cartilage. Risks of surgery include facial nerve damage, decreased hearing, and damage to the vestibular system in the surgical ear. PG decided to undergo surgery, and following the surgical procedure she reported that her symptoms improved by 98%

38.9 Outcome

A definitive diagnosis was initially delayed due to poor quality of the original CT scan; however, the case history and clinical findings clearly suggested SSCD at the time of the initial examination. When PG returned 3 years later, the increased air–bone gap on the audiogram suggested an increase in the size of the dehiscence. Additionally, PG described a notable increase in symptom severity that negatively impacted her quality of life.

38.10 Key Points

1. A CT scan has been considered the gold standard for diagnosis of SSCD; however, a careful case history and audiological findings are also essential in the identification of the disorder. PG's case history and cVEMP were positive for SSCD long before confirmation was made with the repeat CT scan.
2. In the case of conflicting clinical results, further investigation is warranted to reach a definitive diagnosis. Part of the role of the audiologist within the vestibular team is to explain vestibular test findings and be an advocate for the patient and for additional testing, if necessary.

Suggested Reading

Chien W, Ravicz ME, Rosowski JJ, Merchant SN. Measurements of human middle- and inner-ear mechanics with dehiscence of the superior semicircular canal. Otol Neurotol 2007; 28: 250–257

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39 Labyrinthine Fistula

Amy K. Winston and Patricia McCarthy

A 43-year-old female reporting episodic vertigo, hyperacusis, and pulsatile tinnitus in her left ear was referred for an audiological examination.

39.1 Clinical History and Description

BK is a 43-year-old female referred for audiological examination by her primary care physician. She reports severe autophony, hyperacusis, and pulsatile tinnitus in her left ear, which have become more severe over the last 12 to 18 months. She reports she is experiencing brief episodes of intense "true" spinning vertigo that began several years ago and have occurred more frequently over the last year. When asked about what triggers her vertiginous episodes, BK reports that loud sounds, and at times her voice, trigger her vertigo. She has also become very vertiginous while straining, such as when lifting heavy items or having a bowel movement. As a final note, BK states she recently noticed she can "hear" her eyes move in her head.

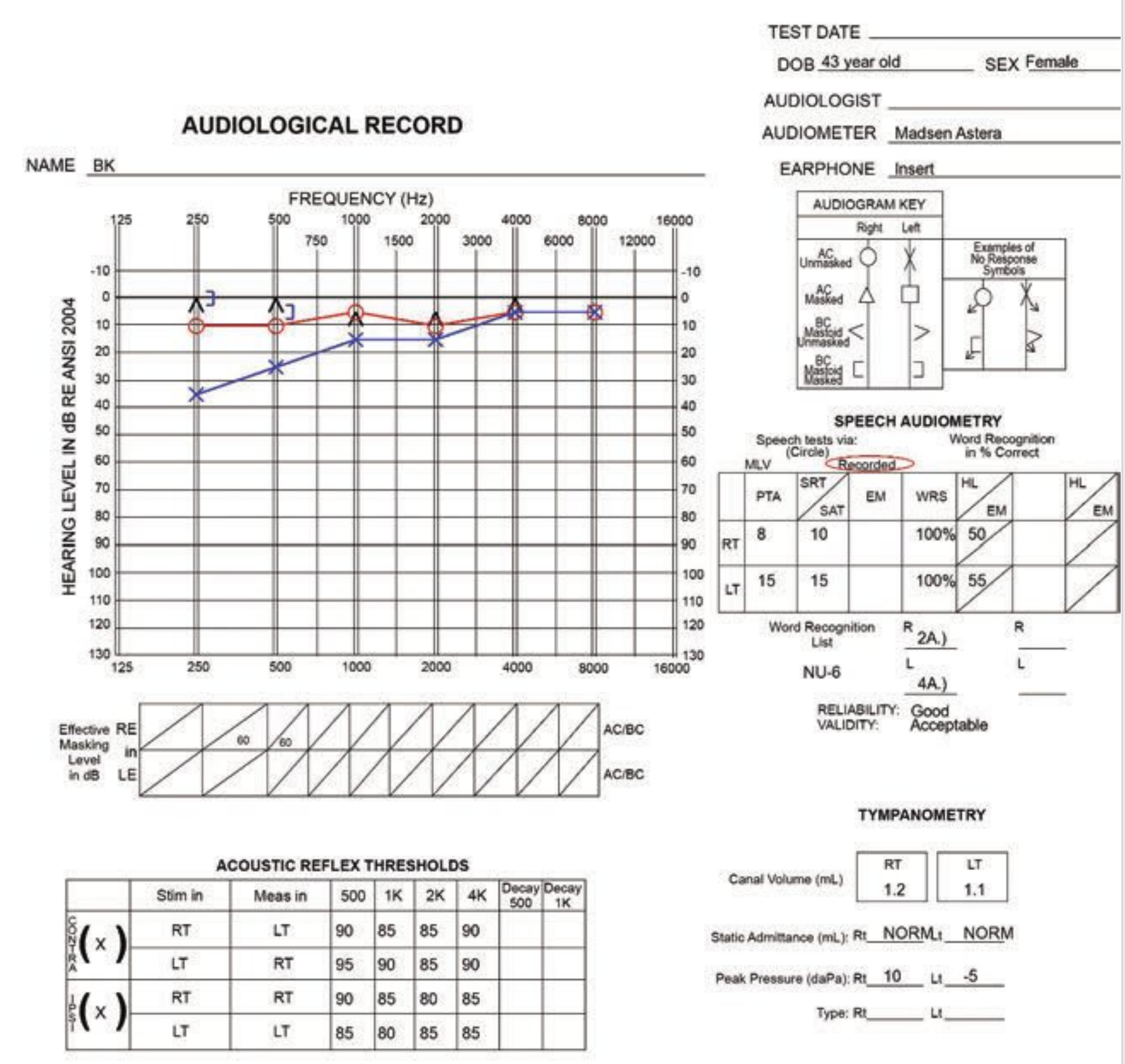
39.2 Audiological Testing

Findings from BK's audiological examination are reported in Fig. 39.1. Pure-tone findings indicated hearing thresholds are within normal limits 250 to 8000 Hz in the right ear, and results for the left ear revealed a mild conductive loss at 250 to 500 Hz rising to within normal limits at 1000 to 8000 Hz. Speech recognition thresholds (SRTs) indicated normal ability to receive speech bilaterally, and word recognition scores (WRSs) revealed normal ability to recognize speech presented at 50 and 55 dB HL, respectively, for the right and left ear. Tympanometric findings for ear canal volume (mL), middle ear pressure (daPa), and static admittance (mL) were within normal limits bilaterally. It is important to note that BK reported that she became "slightly dizzy" during tympanometric testing of the left ear. Acoustic reflex thresholds (ARTs) were present at normal sensation levels bilaterally at 500 to 4000 Hz for contralateral and ipsilateral presentation. Again, BK reported dizziness during presentation of the contralateral and ipsilateral stimuli to the left ear. At certain points during these measures, BK seemed to tilt slightly to the left at the onset of the stimulus.

A review of BK's audiological and immittance results reveals unexpected findings. Specifically, the presence of normal ARTs with the documented normal tympanometric results and air–bone gaps at 250 to 500 Hz in the left ear is highly unusual. This unique pattern of results was highlighted in the report to the referring physician, and a neurotology consult was recommended.

39.3 Neurotology Evaluation

BK was subsequently referred by her primary care physician to a neurotologist. During the evaluation, the neurotologist videotaped BK pinching her nose as she performed a Valsalva maneuver. Review of the video revealed that this change in pressure



AUDIOLOGICAL RECORD

NAME BK

TEST DATE
DOB 43 year old SEX Female
AUDIOLOGIST
AUDIOMETER Madsen Astera
EARPHONE Insert

Effective Masking Level in dB

RE in		60	60									AC/BC
LE												AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	8	10		100%	50		
LT	15	15		100%	55		

Word Recognition List NU-6
R 2A.) R
L 4A.) L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

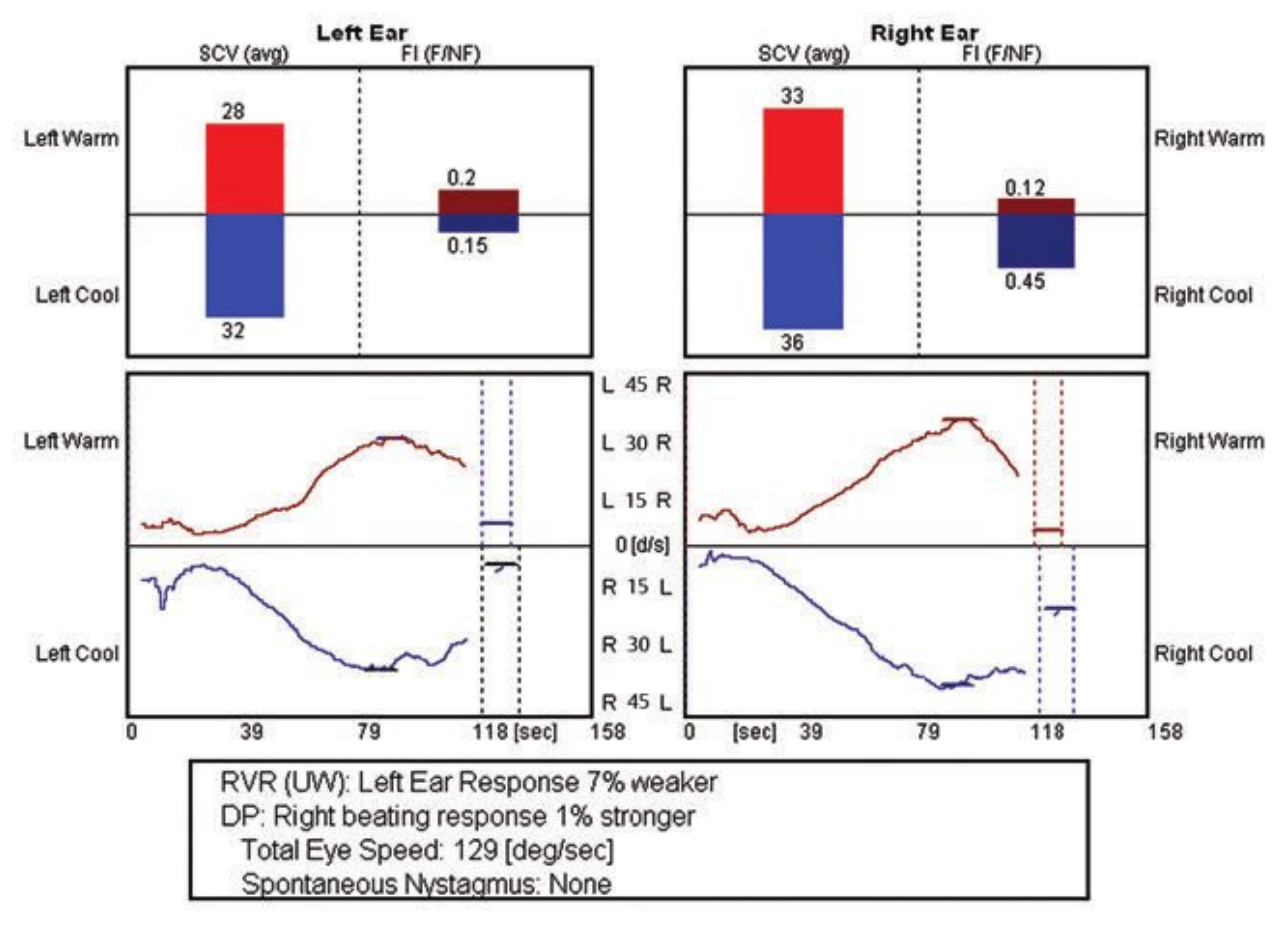
	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	90	85	85	90		
	LT	RT	95	90	85	90		
IPSI (x)	RT	RT	90	85	80	85		
	LT	LT	85	80	85	85		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.1

Static Admittance (mL): Rt NORM Lt NORM
Peak Pressure (daPa): Rt 10 Lt -5
Type: Rt Lt

Fig. 39.1 Audiological examination findings.



RVR (UW): Left Ear Response 7% weaker
DP: Right beating response 1% stronger
Total Eye Speed: 129 [deg/sec]
Spontaneous Nystagmus: None

Fig. 39.2 Bithermal caloric summary.

made BK extremely vertiginous and evoked a downbeating torsional nystagmus, with the fast-phase eye movement rotating in a clockwise direction. BK was referred to return to audiology for videonystagmography (VNG) and cervical vestibular-evoked myogenic potential (cVEMP) examination.

39.4 Questions for the Reader

1. What are the most common causes of sound- and pressure-induced vertigo?
2. Is the direction of BK's nystagmus significant? What information does this provide?

39.5 Discussion of Questions to the Reader

1. What are the most common causes of sound- and pressure-induced vertigo?
   Vertigo caused by loud sounds, known as the Tullio phenomenon, and pressure-related vertigo, known as the Hennebert sign, are unique audiovestibular symptoms. The presentation of these two symptoms suggests a limited range of possibilities in differential diagnosis. A brief list of the most likely etiologies includes Ménière's disease, perilymphatic fistula, and superior semicircular canal dehiscence (SSCD).
2. Is the direction of BK's nystagmus significant? What information does this provide?
   The three semicircular canals (SCCs) are sensitive to angular acceleration within their respective planes of orientation, and through the vestibulo-ocular reflex (VOR), the SCCs trigger reflexive compensatory eye movements within that same plane of action. This relationship is captured succinctly in Ewald's first law: Eye and head movements occur in the plane of the SCC that is being stimulated. Given this, the direction of BK's nystagmus provides insight into which SCC is likely involved. In this case, the neurotologist observed a downbeating and rotary nystagmus in response to a nose-pinch Valsalva, which is an excitatory stimulus. This reaction to the nose-pinch Valsalva suggests vertical canal involvement. In this case, the superior SCC is likely involved.

39.6 Additional Testing

39.6.1 Videonystagmography

VNG test results were normal. No spontaneous, gaze, positional, or post-headshake nystagmus was noted. Findings from oculomotor studies, including gaze testing, smooth pursuit, random saccades, and optokinetic tests, were within normal limits. The Dix-Hallpike yielded normal results in the right and left test conditions. As reported in Fig. 39.2, bithermal caloric testing evoked robust and symmetrical nystagmus in the four test conditions with no significant difference between the ears (unilateral weakness [UW]) and no significant velocity difference when considering the direction of the nystagmus (directional preponderance [DP]).

Cervical Vestibular-Evoked Myogenic Potential (cVEMP)

As reported in Fig. 39.3, cVEMP testing with a 500 Hz tone-burst stimulus revealed a normal cVEMP response for the right and left sides. The cVEMP amplitude, which is measured as the amplitude between the two main peaks of the response (P1 and N1), was within normal limits bilaterally. Latency measures, which are typically 13 to 15 ms for P1 and 21 to 24 ms for N1, were also normal. Note that in this cVEMP result, the initial peak (P1) is a negative or downward-deflecting peak, and the second wave component (N1) is a positive or upward-deflecting peak. It is possible to see cVEMP results with these peaks reversed such that P1 is a positive peak and N1 is a negative peak. This difference in peak direction is the result of the position of the active (noninverting) electrode relative to the reference electrode during recording; it does not alter the interpretation of the cVEMP. These findings were used to calculate a cVEMP asymmetry ratio. The asymmetry ratio (AR) is calculated

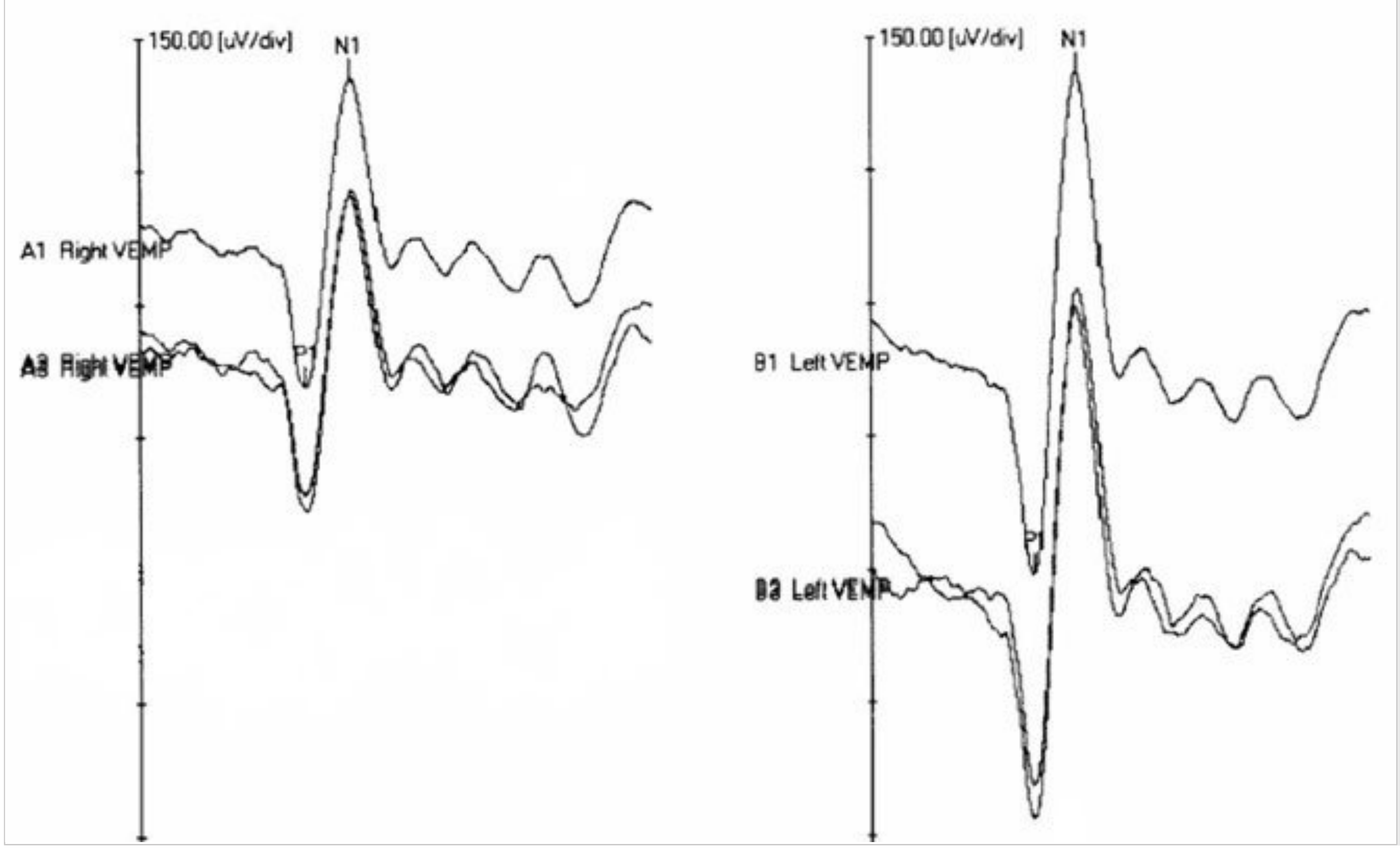


Fig. 39.3 Cervical vestibular-evoked myogenic potential examination findings.

as the amplitude measure for one ear minus that of the opposite ear, divided by the sum of the amplitudes for both ears. In this case, the measured cVEMP amplitude was larger on the left side than the right side, but the calculated AR was within normal limits at 23%

39.7 Additional Questions for the Reader

1. Are the patient's immittance results consistent with the unilateral conductive hearing loss identified at 250 and 500 Hz for the left ear in the audiogram in Fig. 39.1?
2. How is it possible to have a normal caloric response and an abnormal cVEMP?
3. What is significant about the cVEMP findings?

39.8 Additional Discussion of Questions to the Reader

1. Are the patient's immittance results consistent with the unilateral conductive hearing loss identified at 250 Hz and 500 Hz in the audiogram in Fig. 39.1 in the pure-tone testing?
   No, it is unusual to find normal tympanometric and ARTs findings in the presence of a significant conductive component on pure-tone audiometry. Air–bone gaps typically indicate the presence of a middle ear pathology that decreases the efficiency of the middle ear transfer function. With the presence of an air–bone gap, most middle ear disorders result in an abnormal tympanogram and elevated/absent ARTs. Given the presence of air–bone gaps at 250 to 500 Hz in the left ear, an audiologist would have expected an abnormal left tympanogram and elevated or absent ARTs for all ART measures with the exception of the right ipsilateral condition. Some research indicates that tympanometric findings in otosclerosis can be normal depending on the progression and precise site of the disease process. Thus a normal tympanogram would not be inconsistent with a low-frequency air–bone gap. However, the presence of ARTs in all test conditions is inconsistent with otosclerosis. These ART findings suggest that BK's "conductive" component may be a result of something other than a middle ear pathology.
2. How is it possible to have a normal caloric response and an abnormal cVEMP?
   The primary pathway of the cVEMP response is from the saccule to the lateral vestibular nucleus via the inferior branch of the vestibular portion of the vestibulocochlear nerve (cranial nerve VIII). The pathway then continues through the lateral vestibulospinal tract to cranial nerve XI (the accessory nerve), which innervates the muscles of the neck, including the sternocleidomastoid muscle, the primary recording site for the cVEMP. In contrast, bithermal caloric irrigation measures the function of the horizontal SCC, which is innervated by the superior branch of cranial nerve VIII. Because the end organs and vestibular nerve branches investigated by the caloric response and cVEMP are different, it is possible to report a normal finding on one test and an abnormal finding on the other. The cVEMP provides a method to investigate saccular function and the integrity of the inferior branch of the vestibular portion of cranial nerve VIII and therefore makes the cVEMP a valuable addition to a vestibular assessment test battery.
3. What is significant about the cVEMP findings?
   Given the finding of an apparent conductive component in the left ear, the presence of a cVEMP response is diagnostically significant. Research has reported that the intensity of the evoking stimulus is positively correlated with cVEMP amplitude. Although the cVEMP response is not altered by the presence of a sensorineural hearing loss, a middle ear pathology resulting in an air–bone gap can reduce the magnitude of the stimulus reaching the saccule to such an extent that the cVEMP response is abolished.

39.9 Diagnosis and Recommended Treatment

Based on the results of the audiological examination, the examination by the neurotologist, and the findings for caloric and cVEMP testing, the patient was referred for a computed tomographic (CT) scan. Results revealed a dehiscence of BK's left superior SCC.

SSCD is a disorder characterized by an absence of bone overlying the superior canal. The absence of bone overlying BK's left

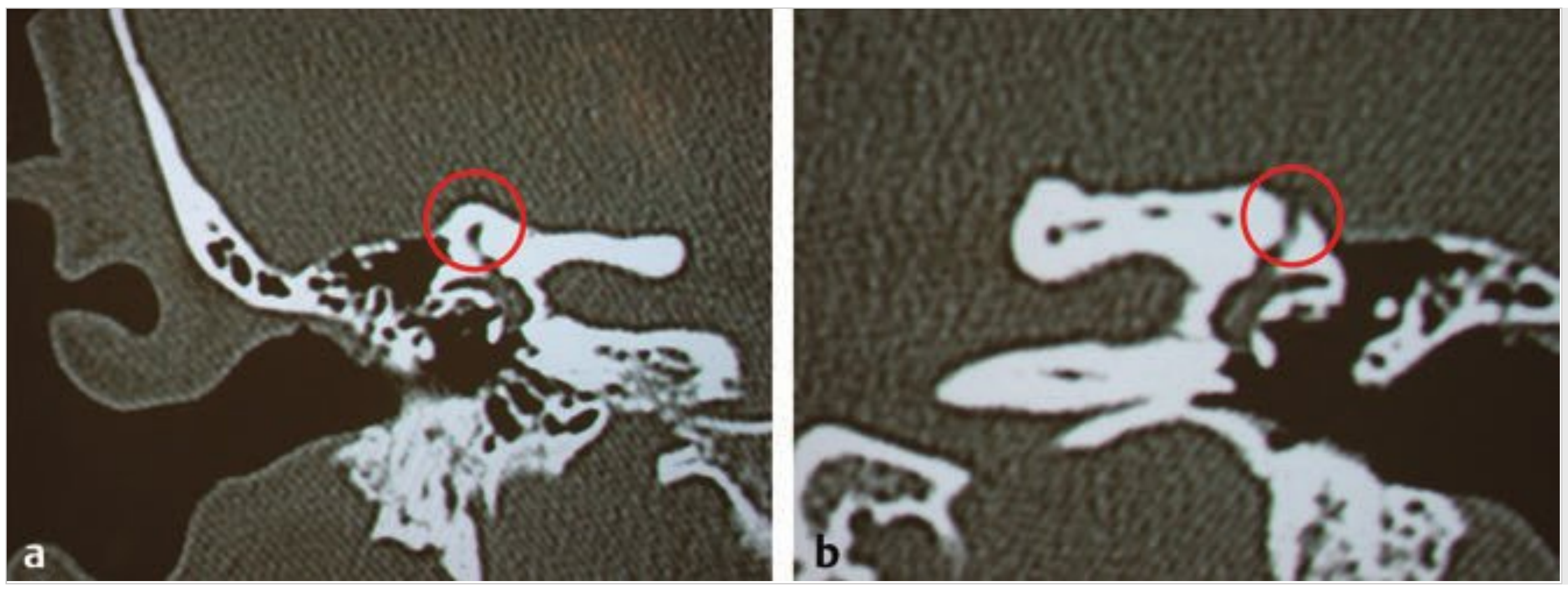


Fig. 39.4 Computed tomographic (CT) scan of the normal right superior semicircular canal (SCC) (a) and the left superior SCC revealing dehiscence (b).

superior SCC is evident in comparing the circled areas in Fig. 39.4. In Fig. 39.4 a, which shows BK's right superior SCC, the bone overlying the canal is clearly intact. In contrast, a large opening in the bone at the top of the left superior SCC can be seen in Fig. 39.4 b. This disorder has been associated with a number of specific otologic signs and symptoms, including sound- and pressure-induced vertigo and nystagmus, air–bone gaps on pure-tone audiometry, vertical oscillopsia (i.e., a perceived up and down movement of the visual field with head movement), autophony, pulsatile tinnitus, persistent imbalance, and hyperacusis. The dehiscence in the bony cover over the superior SCC creates an abnormal pressure outlet in the labyrinthine system, and researchers believe that the constellation of auditory and vestibular symptoms associated with this disorder is directly related to the resulting increase in compliance.

Although the CT scan is the gold standard for differential diagnosis of SSCD, other tests provide important diagnostic details. These include tympanometry; ARTs to ipsilateral and contralateral stimulation; pure-tone audiological examination; tests for sound- and pressure-induced vertigo and nystagmus, including fistula, Valsalva, and Tullio tests; and cVEMP. In some cases of SSCD, the mere presence of the cVEMP response can provide clinicians important diagnostic information. The apparent air–bone gap seen in some SSCD patients is believed to be the result of increased compliance in the labyrinthine system rather than "true" middle ear pathology. Although the precise mechanisms are unclear, presence of the dehiscence seems, at least in some patients, to increase patient sensitivity to bone-conducted sounds, thus reducing bone conduction thresholds, and/or to reduce the efficiency with which low-frequency air-conducted stimuli are transmitted through the middle ear, resulting in elevation of air-conduction thresholds. As a result, the cVEMP response is retained despite the appearance of a conductive component on pure-tone audiometry. It is for this reason as well that the measured ART is retained despite the apparent air–bone gap found in pure-tone audiological testing. In addition, researchers believe that the additional compliance within the labyrinthine system increases the sensitivity of the saccular macula to changes in pressure. Experimental evidence indicates that this increased sensitivity of the vestibular system to pressure changes in cases of SSCD results in alteration of the cVEMP response. Specifically, many researchers have documented findings of augmented cVEMP amplitudes and thus significantly reduced thresholds (the lowest stimulus intensity at which a cVEMP response is present) in the affected ears of subjects with SSCD.

Treatment for SSCD typically involves surgery to plug or resurface the abnormal opening in the superior canal. Resurfacing the dehiscence retains the normal function of the superior canal. In contrast, the plugging approach abolishes the response from the involved canal and triggers the vestibular compensation process, which occurs over a period of weeks to months. In cases where symptoms are mild, a more conservative, watchful approach might be taken in which the patient is educated on how best to avoid triggering conditions.

39.10 Outcome

BK underwent a plugging procedure for her left SSCD and received vestibular therapy to facilitate compensation. When seen by the neurotologist 3 months after her surgery, BK reported a marked improvement in her vertigo, hyperacusis, and pulsatile tinnitus.

39.11 Key Points

1. SSCD is a disorder characterized by an absence of bone overlying the superior SCC.
2. Characteristic symptoms of SSCD include sound- and pressure-induced vertigo, pulsatile tinnitus, hyperacusis, autophony, and oscillopsia.
3. Audiological testing in cases of SSCD may present with an unusual pattern of findings, including an apparent low-frequency air–bone gap with normal tympanometric findings and intact ARTs in the involved ear.
4. Cervical VEMP testing in SSCD cases will show an intact response in the involved ear, despite the apparent conductive component in that ear. In these cases, patients will typically have a large cVEMP amplitude and reduced VEMP threshold relative to the uninvolved ear.

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40 Central Vestibular Dysfunction—Type I Chiari Malformation

Amy K. Winston and Patricia McCarthy

A 30-year-old male was referred by a neurotologist for audiological and videonystagmography (VNG) examinations to investigate patient reports of episodic dizziness following exertion.

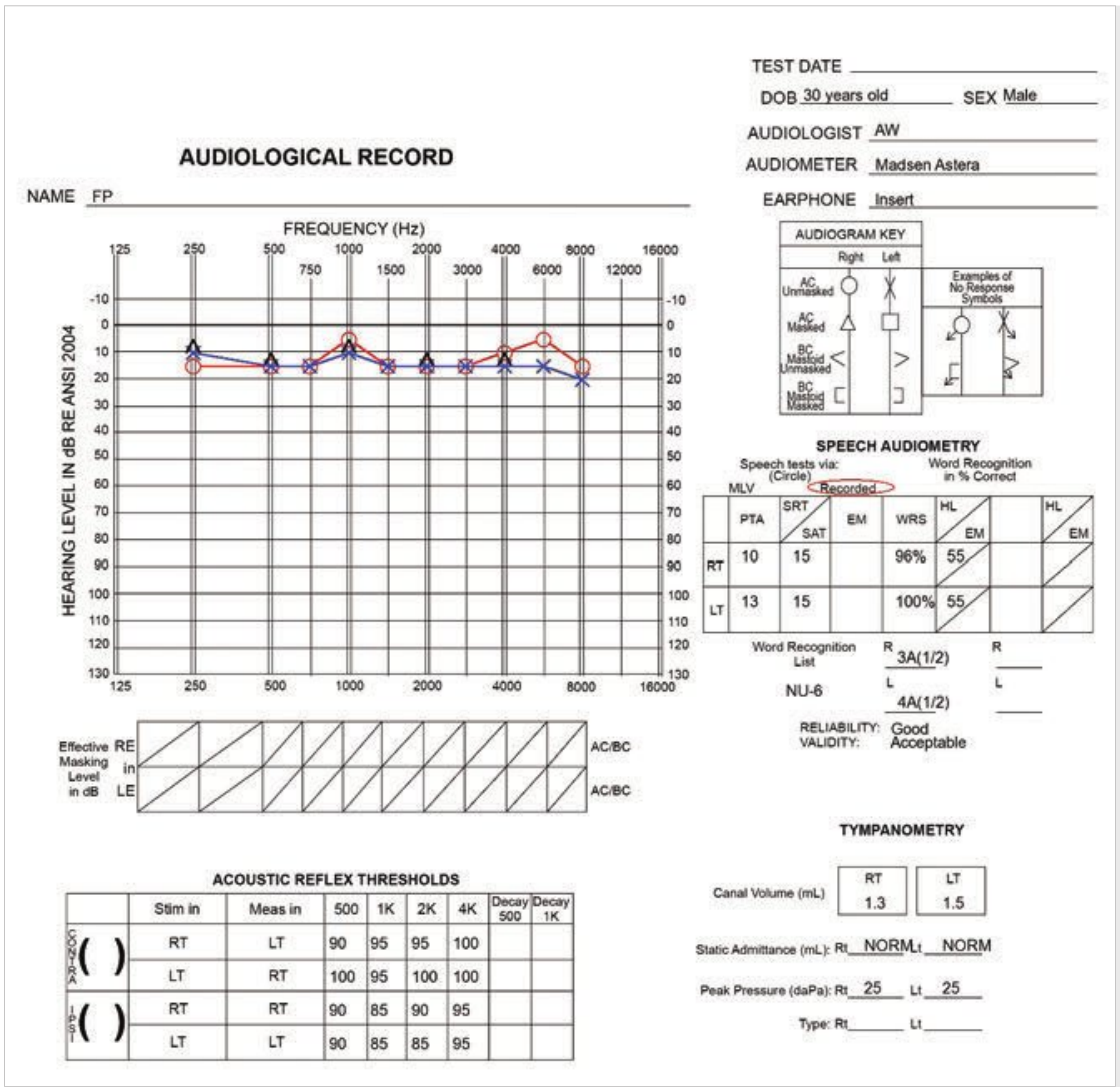
40.1 Clinical History and Description

FP, a 30-year-old male, is referred for audiological and VNG examinations. He reports multiple daily episodes of dizziness lasting less than 30 seconds and most often occurring after some degree of exertion, such as exercising or cleaning the house. When describing the dizziness, FP confirms there is no perception of rotation, either of himself or of his environment. He reports his head is “swimming” and he is unable to focus. There is no accompanying nausea or vomiting, and he denies any hearing concerns and any perceptible changes in hearing during his episodes of dizziness. FP states he first noticed his dizziness almost 3 years ago, but he has recently become concerned because the “dizzy spells” are now occurring several times a day. FP states this problem is beginning to interfere with his quality of life, which is what prompted his visit to his physician.

FP confirms he is free of dizziness between spells, but states he has recently developed visual problems that are concerning. An ophthalmology examination confirmed that his contact lens prescription is appropriate, but FP reports his vision is very blurry during exercise or when he turns his head quickly. Also, he reports he has experienced at least two episodes of diplopia (double vision). In fact, he reports he stopped playing softball because he saw two softballs coming toward him in his last game. FP is also bothered by recurrent headaches that are triggered or exacerbated by sneezing and coughing. These headaches begin at the back of the head and then move down his neck. FP has had headaches since high school, but notes the headaches have become more intense and frequent within the last 6 months. He says he has never been diagnosed with migraine headaches and has never used anything other than over-the-counter pain medication for treatment. True relief from the headaches occurs by lying on his stomach in bed with his head hanging over the edge.

40.2 Audiological Testing

Findings from FP’s audiological examination are shown in Fig. 40.1. Pure-tone hearing thresholds were within normal limits 250 to 8000 Hz bilaterally. Speech recognition thresholds (SRTs) revealed normal ability to receive speech bilaterally and were consistent with the three-frequency pure-tone average. As expected, word recognition scores (WRSs) indicated normal



AUDIOLOGICAL RECORD

NAME FP

TEST DATE

DOB 30 years old SEX Male

AUDIOLOGIST AW

AUDIOMETER Madsen Astera

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	10	15		96%	55		
LT	13	15		100%	55		

Word Recognition List NU-6 R 3A(1/2) L 4A(1/2) R L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	90	95	95	100		
	LT	RT	100	95	100	100		
IPSI ( )	RT	RT	90	85	90	95		
	LT	LT	90	85	85	95		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.3	1.5

Static Admittance (mL): Rt NORM Lt NORM

Peak Pressure (daPa): Rt 25 Lt 25

Type: Rt Lt

Fig. 40.1 Audiological examination findings.

ability to recognize speech bilaterally with the presentation level at a normal conversational level (i.e., 55 dB HL). Tympanometric findings for ear canal volume, (mL) middle ear pressure (daPa), and static admittance (mL) were within normal limits bilaterally. Finally, acoustic reflex thresholds (ARTs) were present at normal or slightly elevated sensation levels relative to pure-tone thresholds at 500 to 4000 Hz.

40.2.1 Dizziness Handicap Inventory

The dizziness handicap inventory (DHI) was administered prior to vestibular testing. The DHI is a self-report questionnaire designed to assess a patient's perception of and emotional response to dizziness, as well as any resulting functional limitations. A total composite score on the DHI provides a measure of the patient's overall level of perceived handicap. On a scale of 0 to 100, with 0 being no perceived handicap and 100 the maximum handicap, a composite score of 16 to 34 is interpreted as a mild handicap, 36 to 52 is a moderate handicap, and 54 or greater is a severe handicap. FP's score of 38 confirms that he perceives a moderate handicap due to his dizziness.

40.2.2 Videonystagmography

VNG examination results were abnormal. Significant findings were noted as follows:

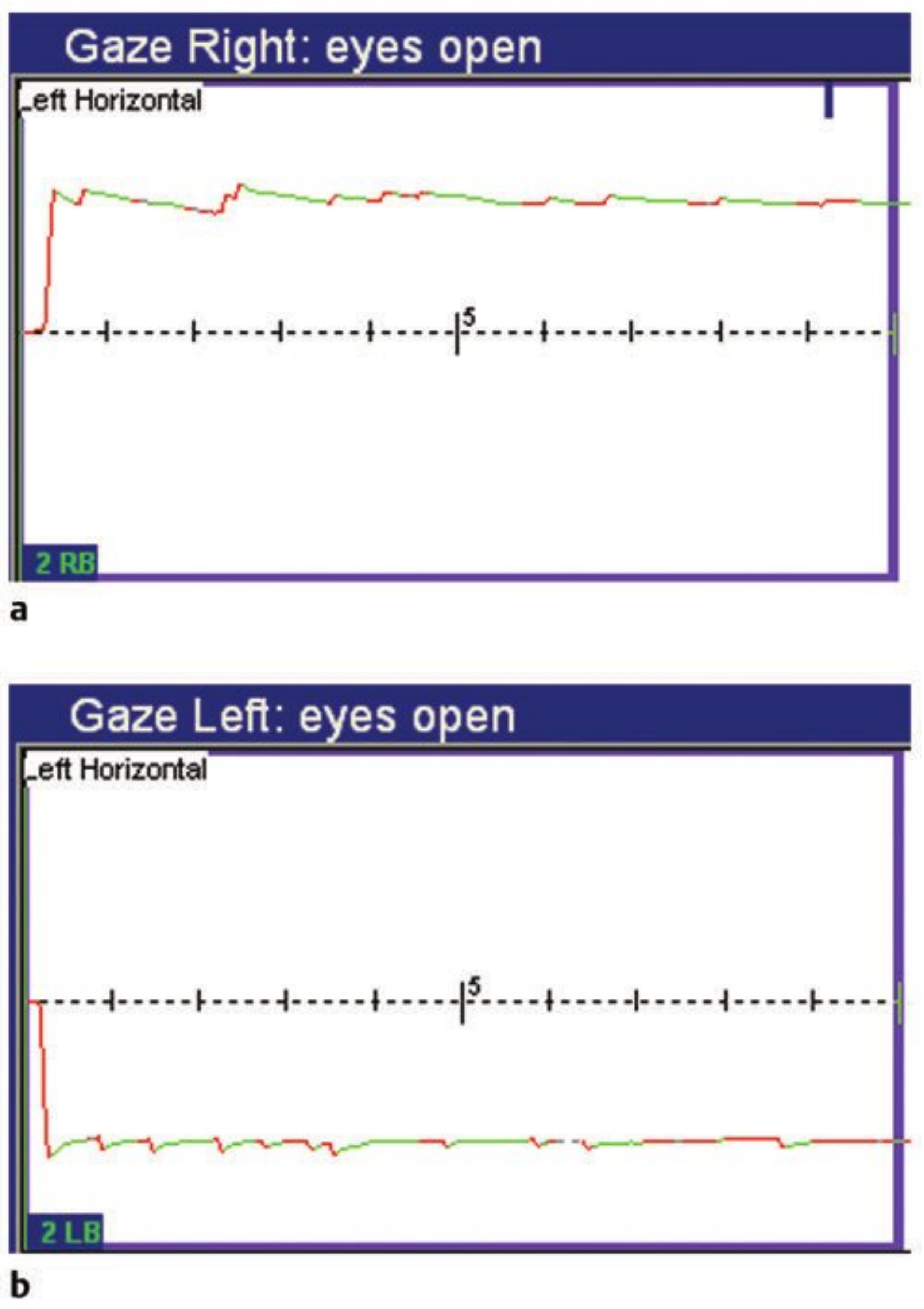


Fig. 40.2 Abnormal gaze test findings revealing a direction-changing gaze nystagmus with fixation consistent with bilateral gaze nystagmus.

a) Gaze testing: A bilateral horizontal gaze nystagmus was identified, with a low-velocity right-beating nystagmus in the gaze right condition with fixation and a low-velocity left-beating nystagmus in the gaze left condition with fixation (Fig. 40.2). In both conditions, nystagmus was absent without fixation.
b) Positional testing: Nystagmus was present in all test conditions, with exacerbations in the conditions without fixation. The nystagmus was direction-changing across different positions, with right-beating nystagmus in the head right and body right conditions and left-beating nystagmus in the head left and body left conditions. Nystagmus velocity was greater in the head and body left conditions relative to these tests on the right.
c) Smooth pursuit testing: Test findings (Fig. 40.3) revealed reduced gain at all test frequencies, with results at 0.2 Hz and 0.4 Hz outside the normative range. Nystagmus consistent with the noted bilateral gaze nystagmus intruded on the tracings, particularly at the 0.1 Hz and 0.2 Hz test frequencies.
d) Optokinetic testing: FP had significantly reduced gain at both the 20°/s and 40°/s target velocities (Fig. 40.4).
e) Bithermal caloric testing: FP's bithermal caloric responses (Fig. 40.5a) were robust and symmetrical, with no significant caloric weakness or directional preponderance. As the fixation index (FI) measures in Fig. 40.5c indicate, failure of fixation suppression was noted in the test conditions that evoke a right-beating nystagmus: left cool and right warm irrigations. Tracings from the right cool irrigation (Fig. 40.5b) further illustrate the patient's inability to adequately suppress the caloric-induced nystagmus.

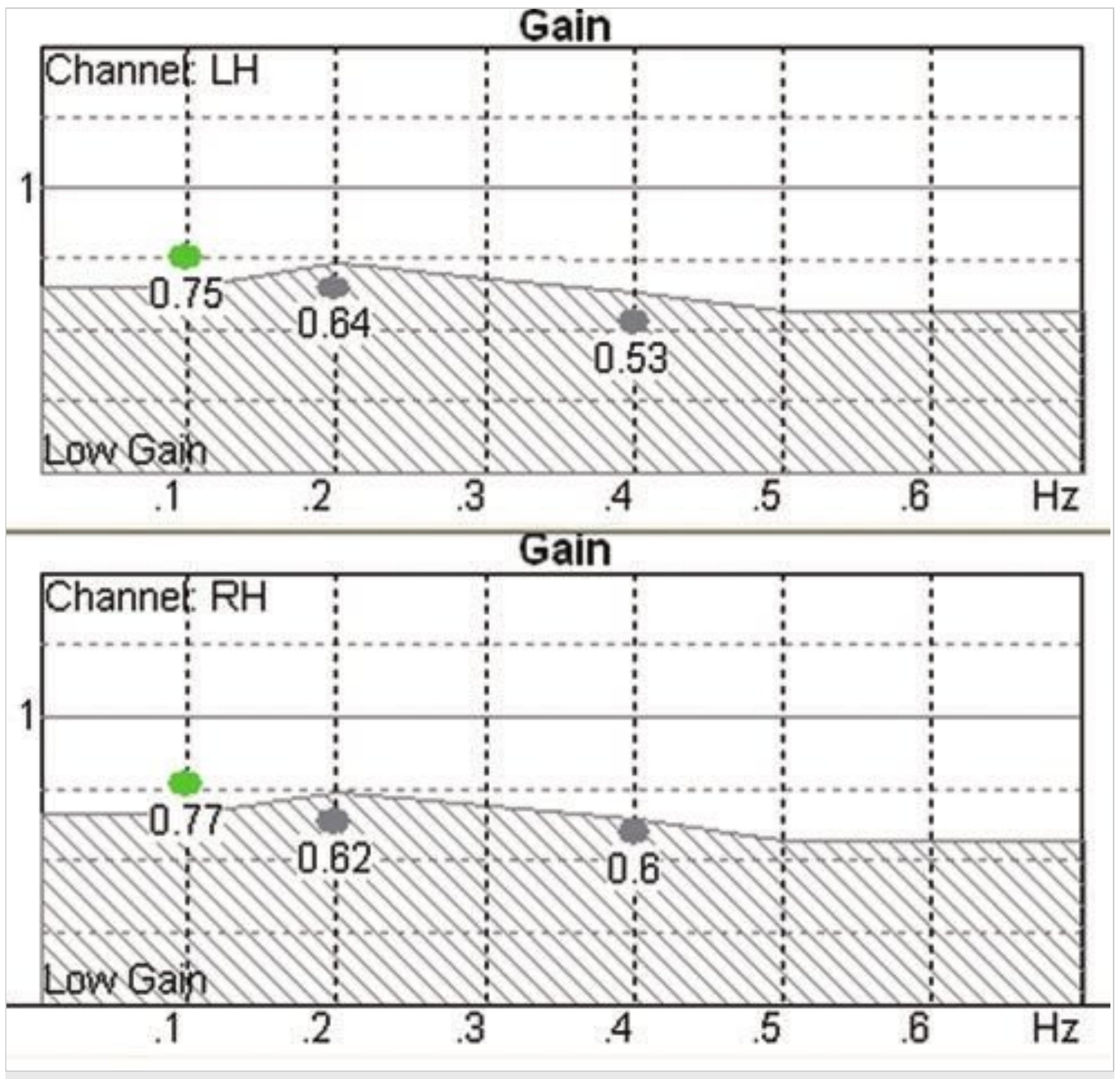


Fig. 40.3 Abnormal smooth pursuit findings indicating reduced gain at the 0.2 Hz and 0.4 Hz test frequencies.

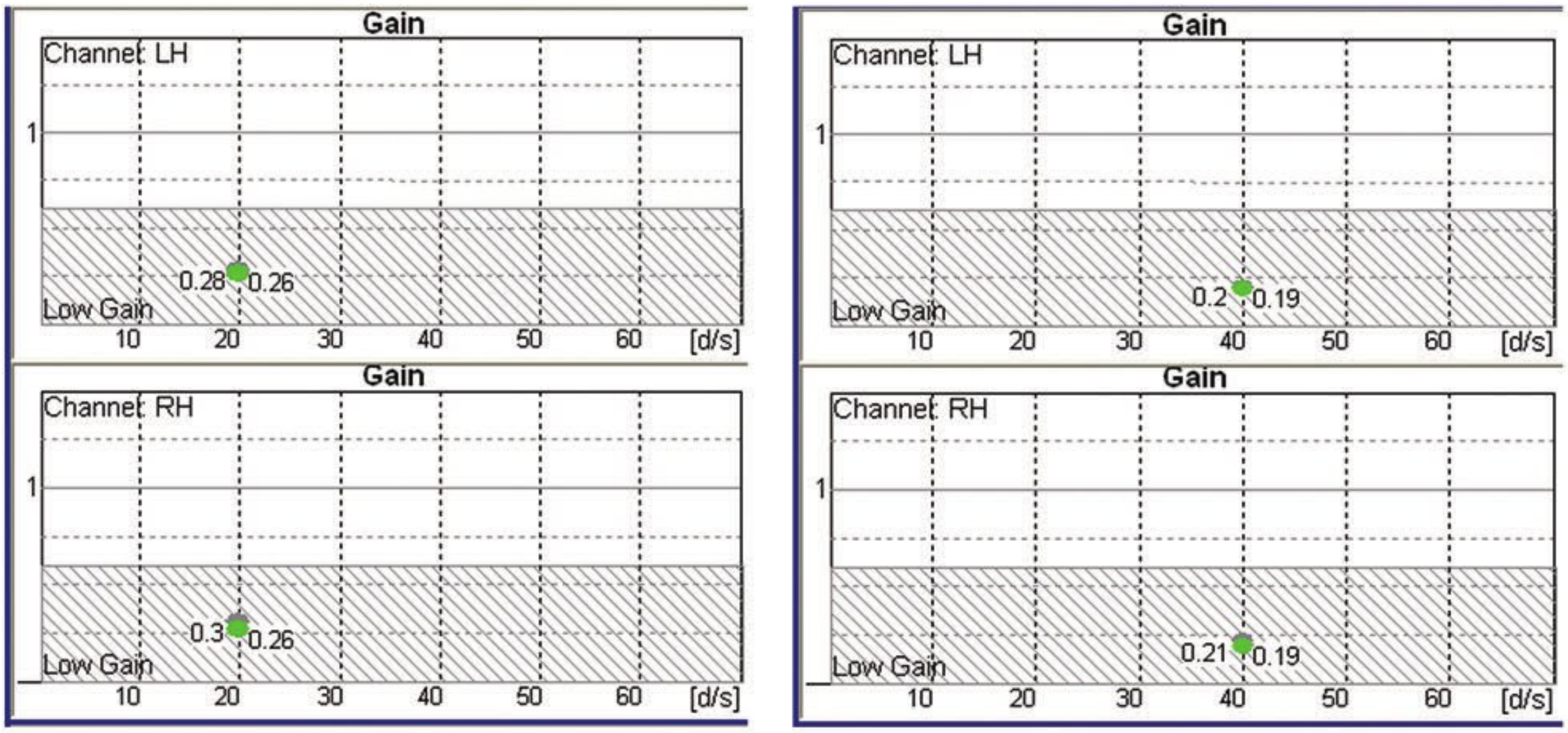


Fig. 40.4 Abnormal optokinetic results with reduced gain at both the 20°/s (OPK 20) and 40°/s (OPK 40) target velocities.

Overall VNG findings suggest that FP's dizziness is central in origin with the cerebellum specifically implicated as a likely site of lesion.

40.3 Questions for the Reader

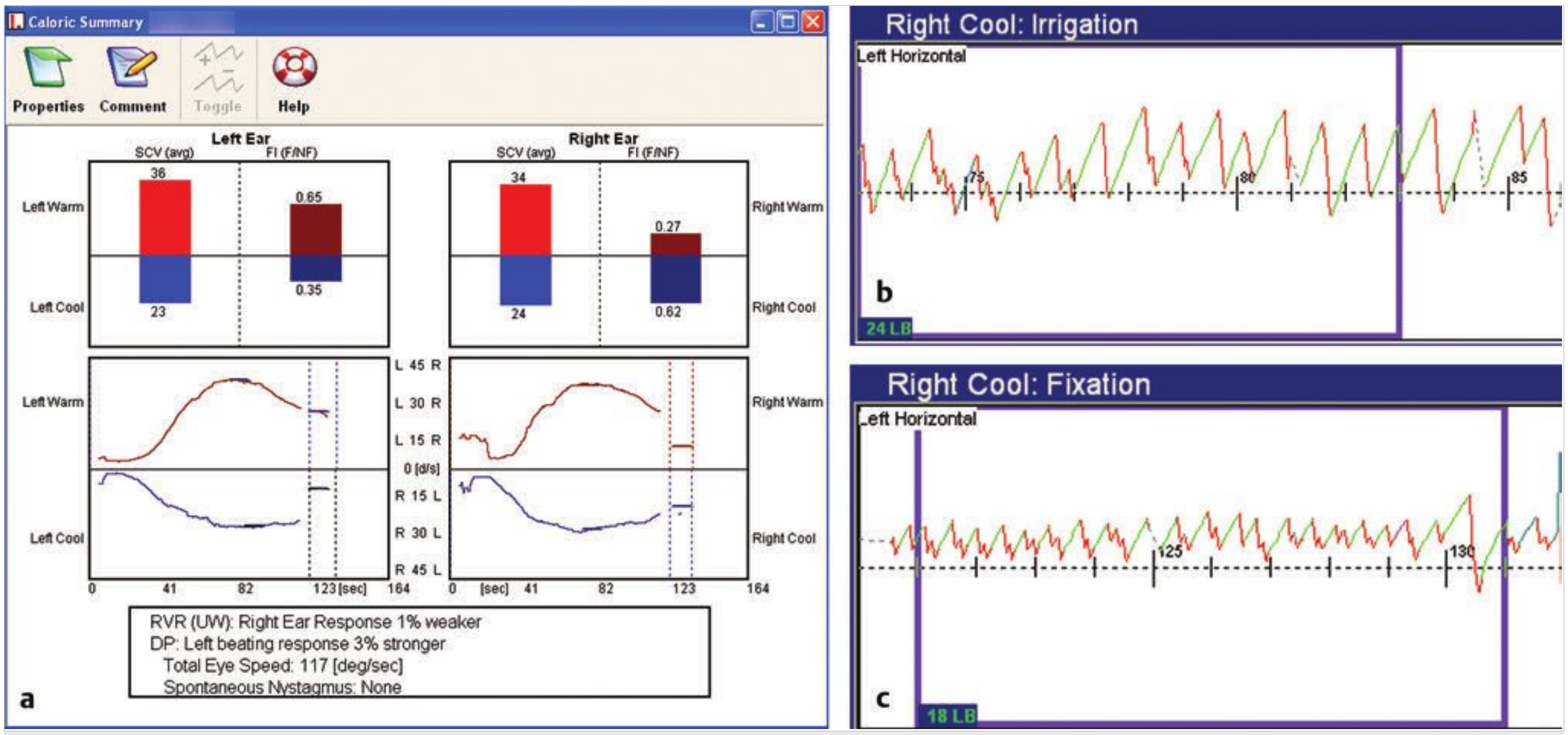
1. Does FP have vertigo?
2. What information is important to consider when taking a vestibular case history?
3. Is there a relationship between migraine headaches and dizziness?

40.4 Discussion of Questions to the Reader

1. Does FP have vertigo?

   Vertigo refers to the false perception of movement of oneself or of one's surrounding environment when no movement is actually occurring. Vertigo may be experienced as a spinning sensation or as a rocking or tilting movement. Based on FP's report of his dizziness, he is not experiencing vertigo.

   Vertigo results from an imbalance in vestibular neural activity, which may originate from peripheral and/or central



RVR (UW): Right Ear Response 1% weaker
DP: Left beating response 3% stronger
Total Eye Speed: 117 [deg/sec]
Spontaneous Nystagmus: None

Fig. 40.5 Bithermal caloric summary (a). Tracings from the right cool caloric irrigation showing peak SPV (b) and fixation (c) as illustration of FP's failure of fixation suppression.

vestibular dysfunction. The three paired semicircular canals are most sensitive to angular acceleration. Unilateral dysfunction in these end organs or their projections will often produce a spinning sensation. Impairment of inputs from the paired otolith organs, the utricle and saccule, which are sensitive to linear acceleration, can generate a feeling of tilting. Other terms that might be used to describe dizziness or a balance problem include lightheadedness, postural imbalance, or disequilibrium. Although the presence of vertigo typically reflects vestibular system dysfunction, nonvertigo dizziness may originate from impairment in other areas of the body, such as the cardiovascular or ocular systems.

2. What information is important to consider when taking a vestibular case history?
   In addition to accurately characterizing the patient's vestibular symptoms as vertiginous or nonvertiginous, there are several other aspects of the patient's experience that can be invaluable in differential diagnosis. Information such as the duration of the dizziness, frequency of occurrence, factors that trigger onset or reduce the intensity of symptoms, associated symptoms (e.g., hearing loss, tinnitus, fullness, etc.), and previously diagnosed conditions (e.g., migraine headache, stroke, skull fracture) can provide important clues about possible site of lesion and causation. For example, episodic vertigo that occurs for only seconds is likely due to peripheral vestibular dysfunction. When triggered by specific changes in head position, this recurrent and brief vertigo is likely indicative of benign paroxysmal positional vertigo. In contrast, longer recurrent vertiginous episodes accompanied by hearing loss, tinnitus, and aural fullness suggest Ménière's disease as a possible cause. Also, a lengthy vertiginous episode lasting 2 to 3 days that is preceded by onset of shingles suggests vestibular neuronitis. In general, vertigo resulting from unilateral peripheral vestibular dysfunction is typically accompanied by nausea and vomiting. These symptoms are typically absent or less severe in vertigo of central origin.
3. Is there a relationship between migraine headaches and dizziness?
   It has been estimated that approximately 30%of individuals with migraines experience migraine-associated vertigo. Migrainous vertigo is not formally recognized as a migraine subtype in the current International Classification of Headache Disorders (ICHD-II), but working criteria for diagnosis of this disorder have been proposed by several investigators. Primary among these criteria is the occurrence of two vertiginous episodes with migraine symptoms, including headache, photophobia (extreme sensitivity to light), and/or phonophobia (extreme sensitivity to sound). Research indicates that vertiginous attacks associated with migraine can take many forms, including spinning, rocking, and intense lightheadedness. Duration of the vertiginous symptoms varies across individuals from a few minutes to several hours. These symptoms can occur concomitant to a headache, prior to the headache (as a vestibular aura), or even in the absence of a headache. In some patients with migraine-associated vertigo, their vertiginous attacks are triggered by the same things known to precipitate their migraine headaches. These may include weather changes, consumption of certain foods such as aged cheeses, and menses.

40.5 Additional Questions to the Reader

1. Can direction-changing gaze nystagmus result from a peripheral vestibulopathy?
2. What is the cause of FP's blurred vision with head movement?
3. What is the significance of failure of fixation suppression?

40.6 Discussion of Additional Questions to the Reader

1. Can direction-changing gaze nystagmus result from a peripheral vestibulopathy?
   There are several important reflex pathways that respond to input from the peripheral vestibular end organs. One of these reflexes, the vestibulo-ocular reflex (VOR), functions to help maintain clear vision during locomotion by generating specific compensatory eye movements in response to perceived head movements. The VOR uses input from the semicircular canals to direct the actions (contraction or relaxation) of corresponding extraocular muscles and thus generate predictable eye movements in the direction opposite that of the perceived head movement. Because they alter the input from the affected semicircular canals to the VOR, unilateral peripheral vestibular disorders generate a predictable, constant-velocity vestibular nystagmus that is of fixed direction regardless of eye position. Nystagmus with a central etiology does not originate from inaccurate peripheral inputs to the VOR and thus may be of fixed direction or may change direction, such as is seen with FP.
2. What is the cause of FP's blurred vision with head movement?
   The VOR, when considered in its simplest form, is a three-neuron reflex arc that functions to assist in the maintenance of clear vision during head movement. Through this reflex arc, information concerning the speed and direction of head movement from the paired semicircular canals is conveyed through Scarpa's ganglion to the vestibular nuclei and then to the appropriate oculomotor nuclei, thus triggering compensatory eye movements. If information from the semicircular canals to this reflex pathway is inaccurate, as might happen with a peripheral vestibular disorder, or if central dysfunction prevents accurate conveyance of this information, an abnormal VOR response results. An abnormal VOR response can lead to oscillopsia, or blurring of vision with head movement, due to inappropriate gain, timing, or direction of the resulting eye movement.
3. What is the significance of failure of fixation suppression?
   Failure of fixation suppression is the inability to adequately suppress caloric-generated nystagmus. The presentation of a visual target triggers the visual pursuit system to generate compensatory eye movements in an attempt to keep the target image on the fovea, resulting in suppression of the caloric nystagmus. Research suggests that fixation suppression is mediated by the cerebellum, specifically the cerebellar flocculus, and that patients with failure of fixation suppression also have impaired visual pursuit systems.

40.7 Diagnosis and Recommended Treatment

Subsequent magnetic resonance imaging (MRI) revealed that FP has a Type I Chiari malformation. Chiari I malformation is a condition in which the cerebellar tonsils and brainstem become displaced from the skull and into the spinal canal, which can result in compression of these tissues and disruption in the normal flow of cerebrospinal fluid. FP's MRI scan indicated that his cerebellar tonsils were displaced 3.5 cm below the foramen magnum and into the spinal canal to the level of C2–C3. Type I Chiari malformation is also known as adult Chiari, because rather than being present at birth, adult Chiari develops as the brain and skull grow. In many cases, individuals with Type I Chiari malformation are asymptomatic or do not become symptomatic until adulthood. Typical symptoms of Type I Chiari malformation include problems with dizziness and balance/coordination, vision changes, headaches, and muscle weakness.

Depending on the severity of symptoms, treatment for Type I Chiari malformation can vary from observation to surgery. FP was scheduled for Chiari revision surgery, which includes suboccipital decompression and C1–C2 laminectomy—removal of the back part of the vertebra that covers the spinal canal—within weeks of his VNG testing. The goal of decompression surgery is to create more room in the skull for the cerebellum; the laminectomy effectively enlarges the spinal canal within the surgical area. Research suggests that, in the majority of cases, suboccipital decompression surgery resolves many of the oculomotor and vestibular symptoms of Type I Chiari malformation patients, including oscillopsia, diplopia, and blurred vision. There is evidence to suggest that typical oculomotor test findings on VNG, such as failure of fixation suppression and poor smooth pursuit and optokinetic results, are improved following surgical intervention.

40.8 Outcome

In FP's case, postsurgery evaluation by his neurotologist revealed FP to be free of balance complaints, with no recurrence of his dizziness, diplopia, or blurred vision.

40.9 Key Points

1. Chiari I malformation is a condition in which the cerebellar tonsils and brainstem become displaced from the skull and into the spinal canal, which can result in compression of these tissues and disruption in the normal flow of cerebrospinal fluid.
2. Typical symptoms of Type I Chiari malformation include dizziness, balance/coordination problems, vision changes, headaches, and muscle weakness.
3. Oculomotor abnormalities in VNG testing, including direction-changing gaze nystagmus, reduced gain on smooth pursuit and optokinetic tests, and failure of fixation suppression on caloric testing, are inconsistent with peripheral vestibular dysfunction and suggest and underlying central pathology, with the cerebellum as the likely site of lesion.
4. Research suggests that surgical intervention, including laminectomy and suboccipital decompression, can resolve many of the oculomotor and vestibular symptoms of Type I Chiari patients.

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Part 4

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41 Modifying Insertion Gain without Computer-Based Software

Amyn M. Amlani

This case report demonstrates techniques for modifying gain on a patient's hearing aid when manufacturer software is unavailable.

41.1 Clinical History and Description

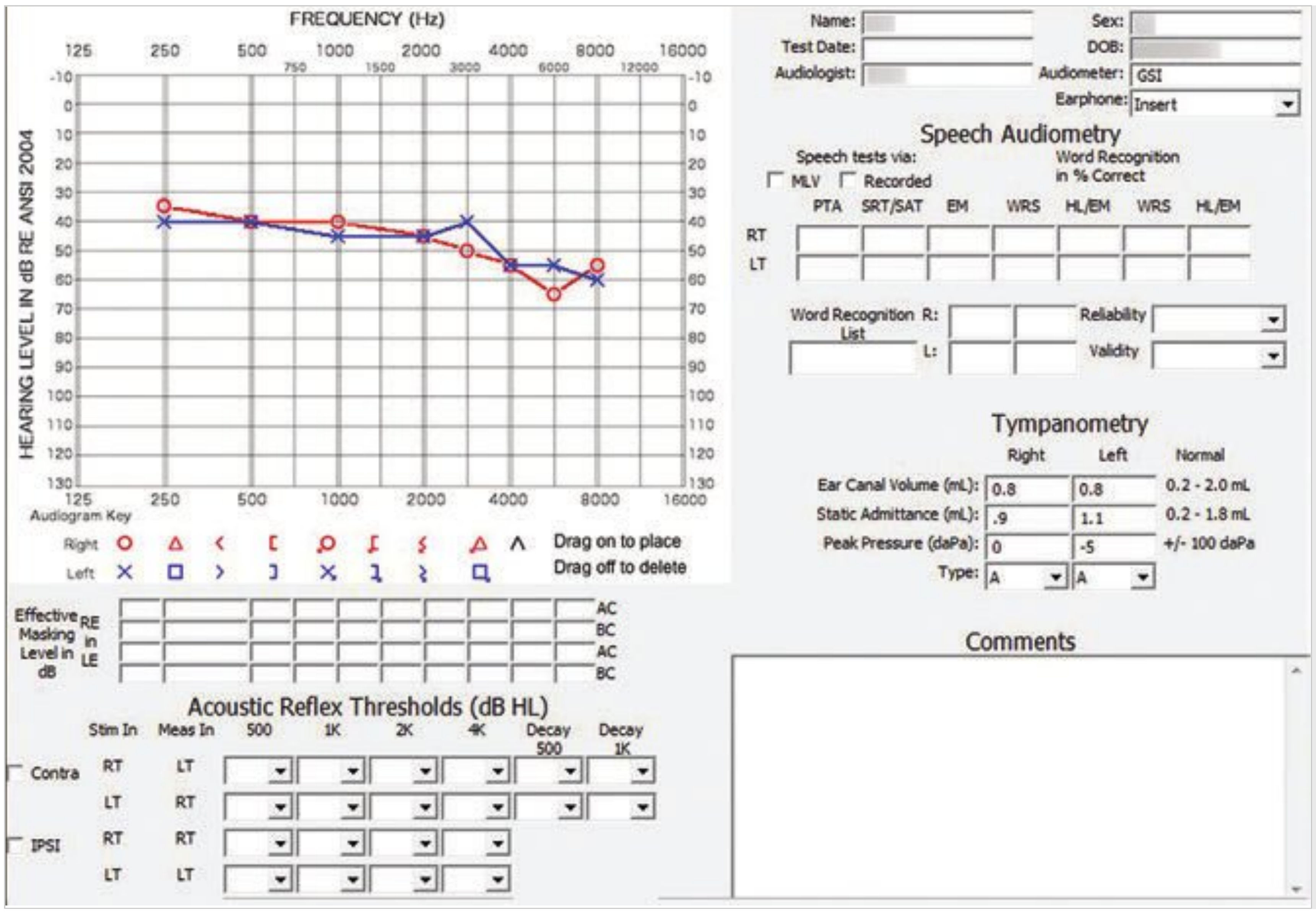
AS is a 51-year-old structural engineer employed by a commercial development company. Her responsibilities include the design, construction, and restoration of commercial buildings across the United States. AS wears bilateral behind-the-ear (BTE) digital hearing devices coupled to custom-made silicone skeleton earmolds. The earmolds include no. 13 standard tubing and a 2 mm parallel vent. AS purchased the hearing devices approximately 18 months ago and recently replaced her old earmolds with these newer earmolds.

AS frequently travels for work. The day she was to depart on an assignment, AS scheduled a routine hearing aid checkup appointment with the practice where she bought the hearing devices. After leaving the practice, AS noted a different sound quality to her hearing devices, but did not have the time to return to the practice for readjustment. That evening, AS, who had traveled to another state, noticed that aided conversational speech in quiet lacked sufficient audibility, Knowing that her hearing devices were the problem, she walked into an audiology practice the following morning and requested an inspection of her hearing devices. The front desk staff informed the patient that the practice did not support this particular manufacturer, but would ask the audiology staff whether they might be able to assist. An audiologist agreed to assist AS, despite the inability to reprogram the hearing devices using a personal computer (PC). The clinician performed an online search of the hearing devices and found the manufacturer's specifications. The specifications indicated that the hearing devices were designed with three channels, each employing wide-dynamic range compression (WDRC).

The clinician first performed a listening check on the hearing devices, which revealed no perceptual abnormalities in aided sound quality. Then the hearing devices were placed in a hearing aid test box to rule out distortion and potential abnormalities in gain and output. Because the hearing devices could not be programmed to test mode using the manufacturer's software, 2 cc electroacoustic results were not compared to manufacturer-reported specifications.

Next, the clinician performed an abbreviated audiological examination, which is standard protocol in this clinic for a hearing device adjustment. The abbreviated examination consisted of air conduction thresholds using insert earphones and tympanometry. Audiological results, reported in Fig. 41.1 revealed a mild to moderately severe hearing loss and type A tympanogram. These audiological results suggest a sensorineural hearing loss based on the tympanogram results representing normal middle ear function.

The clinician entered the patient's hearing threshold data into a real-ear analyzer, which calculated target gain using the National Acoustic Laboratories' nonlinear fitting formula versions 1 (NAL-NL1) prescriptive formula for a 65 dB sound pressure level (SPL) input level, three-channel device and binaural fitting. Next, the clinician performed real ear insertion gain (REIG) where REIG = Real ear aided gain (REAG) – Real ear unaided gain (REUG) using a 65 dB SPL input signal. Table 41.1 reports the measured REIG and NAL-NL1 target gain across frequencies for the right ear (for illustrative purposes, only right ear results are reported in this case study; however, all concepts and calculations can be applied to the left ear).



Name: [redacted]
Test Date:
Audiologist: [redacted]
Sex: [redacted]
DOB: [redacted]
Audiometer: GSI
Earphone: Insert

Tympanometry

	Right	Left	Normal
Ear Canal Volume (mL):	0.8	0.8	0.2 - 2.0 mL
Static Admittance (mL):	.9	1.1	0.2 - 1.8 mL
Peak Pressure (daPa):	0	-5	+/- 100 daPa
Type:	A	A	

Fig. 41.1 Audiological results for AS.

Table 41.1 Difference in real ear insertion gain (REIG) between measured REIG and NAL-NL1 target gain at 65 dB sound pressure level for the right ear.

	Frequency (Hz)						
	250	500	1000	2000	3000	4000	6000
Measured REIG	2.6	6.4	8.2	12.1	13.4	14.6	12.9
NAL-NL1 target gain	11.8	16.2	19.0	19.9	21.1	22.7	25.8
Difference	− 9.2	− 9.8	− 10.8	− 7.8	− 7.7	− 8.1	− 12.9

Clearly, there is a substantial difference between the measured REIG and prescribed NAL-NL1 gain provided by the hearing aid. The patient was informed that her inability to hear as well as before stems from inadequate gain provided by the hearing aids.

41.2 Questions to the Reader

1. Can gain be modified in different frequency regions to meet a prescribed target without the availability of the manufacturer's software?
2. What additional factor(s) must be considered prior to modifying gain of the hearing aids?
3. How did the clinician's actions impact the practice?

41.3 Discussion of Questions to the Reader

1. Can gain be modified in different frequency regions to meet a prescribed target without the availability of the manufacturer's software?

   Without the use of computer-based software, the clinician realized his ability to modify gain at different frequency regions was restricted primarily to the acoustical properties of the earmolds. For example, venting primarily influences the low-frequency response (<1000 Hz). An occluded earmold (i.e., no vent) provides a marked increase in aided gain/output below 1000 Hz. Widening the vent diameter reduces the aided gain/output below 1000 Hz in the ear canal because amplified sounds within this frequency range escape through the vent into the ambient environment.

   In addition, the type of vent configuration influences the acoustic signal in the ear canal. A parallel vent is one in which the vent bore is placed below and parallel to the sound bore. In this configuration, only the low-frequency spectra are altered. A diagonal vent, on the other hand, is one in which the vent begins at the lateral surface of the earmold and then intersects the sound bore between the tubing and the earmold at the medial end. Diagonal venting is not recommended because of its adverse effect on reducing gain/output in the high-frequency region, with the greatest reduction in gain/output occurring when the vent intersects the sound bore in a more lateral location. Because the venting in the earmolds is parallel in this case study, changes in the gain/output are restricted to only the low-frequency region.

   Tubing length and internal diameter also influence the acoustic signal in the ear canal, primarily in the high-frequency region. Electroacoustic testing of BTE hearing devices is standardized using no. 13 tubing having a length of 25 mm. Increasing the tube length will result in the primary resonant peak shifting downward and the gain/output increasing in the low-frequency region and decreasing the gain/output in the mid- and high-frequency regions. Shortening tube length results in the primary resonant peak shifting upward and increasing the gain/output in the mid- and high-frequency regions and decreasing gain/output in the low-frequency region.

   The influence of the tubing's internal diameter is dependent on whether the diameter is constant or stepped. No. 13 standard tubing has a constant internal diameter of 1.93 mm. If the constant internal diameter is widened for tubing characteristics having values < no. 13, based on characteristics standardized by the National Association of Earmold Laboratories (NAEL), the primary resonant peak will shift upward, increasing the gain/output in the mid- and high-frequency regions (i.e., > 750 Hz) and slightly decreasing the gain/output in the low-frequency region (i.e., < 750 Hz). If, instead, the constant internal diameter is narrowed (i.e., tubing values > no. 13 per NAEL), the primary resonant peak will shift downward, slightly increasing the gain/output in the low- and mid-frequency regions (i.e., < 1500 Hz) and decreasing the gain/output in the high-frequency region (i.e., > 1500 Hz).

   A limitation of tubing with a constant-bore diameter is that the high-frequency response of the hearing aid may be lowered from 6000 to 4000 Hz. This limitation led to the development of a single-stepped bore horn configuration known as the Libby horn. The 3 mm Libby horn is designed with the initial 21 mm having an internal diameter of 1.93 mm (i.e., no. 13 tubing), then gradually widening to an internal diameter of 3 mm over the remaining 22 mm. Similarly, the 4 mm Libby horn is designed with the initial 21 mm having an internal diameter of 1.93 mm (i.e., no. 13 tubing), which then gradually widens to an internal diameter of 4 mm over the remaining 22 mm. The 3 mm horn provides approximately 8 to 10 dB greater high-frequency gain/output (i.e., > 3000 Hz), whereas the 4 mm horn provides increased gain/output of 10 to 12 dB in the high frequencies (i.e., > 3000 Hz).

   With venting providing options for modifying the low-frequency region and tubing considerations providing modifying options for the high-frequency region, dampers allow for modifying the midfrequency region (i.e., 1000–3000 Hz). Dampers act to smooth resonant peaks in the frequency response and provide a gently rising high-frequency response. There are several types of damping material, with the fused mesh material being the most widely used. In BTE hearing devices, the fused meshed style is usually placed at the tip of the earhook. Dampers are manufactured in a variety of resistances, with 680, 1500, 2200, 3300, and 4700 Ω being the most common. In general, dampers reduce aided gain/output in the midfrequency region, with the greatest reduction (i.e., ~ 20 dB) occurring for a damper having 4700 Ω resistance and

Table 41.2 Individually measured and average real ear to coupler differences (RECDs) for the right ear

	Frequency (Hz)						
	250	500	1000	2000	3000	4000	6000
Measured RECD	4.0	6.0	8.0	10.0	12.5	13.7	11.4
Average RECD	5.0	6.0	8.0	10.0	7.0	10.0	10.0
Difference	− 1.0	0.0	0.0	0.0	5.5	3.7	1.4

the least reduction (i.e., ~ 6 dB) occurring for a damper having 680 Ω resistance.

2. What additional factor(s) must be considered prior to modifying the gain of the hearing aids?

The clinician conceded that two options were available: blindly modify the gain of the hearing and earmold or estimate the modified insertion gain using the coupler response for flat insertion gain (CORFIG). CORFIG is a set of device-specific correction factors used to predict target gain between REIG and measurements made in a 2 cc coupler. CORFIG estimates insertion gain using specific data, such as the patient's ear canal volume and middle ear impedance, hearing aid microphone location, and earmold venting and tubing considerations. Because CORFIG provides for individual correction factors, as opposed to gross non-patient-specific estimates, the clinician decided to use this process.

Estimating REIG target required obtaining the patient's REUG and real ear to coupler difference (RECD), and employing standardized correction values for hearing aid microphone location. These factors must be considered because of their influence on the SPL in the ear canal. Specifically, the insertion of a hearing aid results in a loss in the natural resonance of the unaided ear canal (i.e., REUG). The insertion of the hearing aid also results in a smaller volume than the 2 cc coupler, increasing gain/output. This increase in gain/output is the RECD. In addition, the placement of the microphone deeper into the concha increases gain/output.

The clinician began by obtaining the RECD. The measured RECD provided the clinician with the acoustic contribution of AS's ear canal to the overall SPL present at the tympanic membrane. RECD is the difference in SPL between the 2 cc coupler, the small-volume cavity used during hearing aid electroacoustic analysis, and the patient's ear canal. The coupler component is performed in a hearing aid test box without the presence of the patient. The real-ear component of the RECD, however, requires the careful placement of a probe tube in the patient's ear canal. In most adult patients, the 2 cc coupler will underestimate the true SPL provided by the average human ear canal. This outcome stems from the 2 cc coupler having a larger volume than the average adult ear canal and its inefficiency to reflect individual differences in acoustic impedance of the human ear canal.

Table 41.2 reports the average RECD values, adopted from NAL-NL1, and the individually measured RECD values for AS's right ear. The clinician notes that AS's individual RECD values deviate from the average RECD value, indicating the SPL produced in her ear canal is greater than the SPL measured in a 2 cc coupler. Stated differently, the larger RECD values measured on AS indicated that the patient's ear canals are much smaller in volume than the standard 2 cc coupler volume used in hearing aid electroacoustic analysis. The difference between measured RECD and average RECD differs across patient age, patient gender, and hearing aid insertion depth.

The clinician also obtained REUG data on AS's right ear. This procedure was performed by positioning the loudspeaker at 45° azimuth. The desired 2 cc coupler response (i.e., CORFIG) was determined by subtracting the measured RECD and the predicted BTE hearing aid microphone location response from AS's REUG (i.e., CORFIG = REUG – RECD – Microphone location). This calculation is shown in Table 41.3. From these data, the clinician derived AS's resulting 2 cc coupler measure by summing the measured REIG (Table 41.1) and the individual CORFIG. In this example, the measured REIG is used as opposed to an NAL-NL1 target because of the clinician's inability to adjust gain/output using the manufacturer's software. The resulting 2 cc coupler target is then compared with the NAL-NL1 target. As noted in Table 41.3, the hearing aid provided markedly less gain than the NAL-NL1 target.

The 2 cc coupler target does not account for venting or for tubing that differed from the no. 13 standard tubing. To account for these factors, the clinician accessed these correction factors from a hearing aid textbook. The clinician estimated the increase in gain/output for a vent size that decreased from 2 to 1 mm by subtracting differences between vent sizes. For instance, the 2 mm vent size estimated an 11 dB reduction in gain at 250 Hz, whereas the 1 mm vent size estimated a 5 dB reduction in gain. The reduction in vent size results in a 6 dB increase in gain at this frequency (Table 41.4). Although the clinician was capable of reducing the vent size from 2 mm to fully occluded, doing so would have increased the occlusion effect for AS.

Correction values were also obtained for a 3 mm Libby horn having a 1500 Ω damper and added to the resulting 2 cc coupler response. The predicted changes in gain stemming from reducing vent size and increasing tubing size are added to the individual CORFIG, resulting in the modified individual CORFIG (Table 41.4). The clinician then combined the results of the modified individual CORFIG to the measured REIG, resulting in an adjusted 2 cc target. This is the estimated REIG expected after all modifications have been made. When compared with the NAL-NL1 target, the adjusted 2 cc target in the low frequencies is slightly increased and markedly increased in the high frequencies. In fact, the adjusted 2 cc target at 3000 and 4000 Hz indicated slightly more gain than the NAL-NL1 target. Although this predicted increased gain in the high frequencies appears to provide the listener with too much gain, the clinician is aware of the variability in the predicted measurements.

Armed with the estimated modifications to gain/output, the clinician modified the earmolds. A postmodification REIG was performed to verify changes in gain. Fig. 41.2 reports REIG for the NAL-NL1 target (asterisks), unmodified earmold

Table 41.3 Calculations of coupler response for flat insertion gain (CORFIG) used to determine 2 cc coupler target for AS's right hearing aid, and the resulting difference between 2 cc coupler and NAL-NL1 targets

		Frequency (Hz)						
		250	500	1000	2000	3000	4000	6000
A. Individual CORFIG	Measured REUG	1.5	6.1	8.7	13.5	17.7	20.7	15.6
	Measured RECD	− 4.0	− 6.0	− 8.0	− 10.0	− 12.5	− 13.7	− 11.4
	Predicted microphone effect	− 0.5	− 1.2	− 0.3	− 4.1	− 2.8	− 3.7	− 1.6
	Individual CORFIG	− 3.0	− 1.1	0.4	− 0.6	2.4	3.3	2.6
B. Comparing targets	Measured REIG*	2.6	6.4	8.2	12.1	13.4	14.6	12.9
	Individual CORFIG	− 3.0	− 1.1	0.4	− 0.6	2.4	3.3	2.6
	Resulting 2 cc target	− 0.4	5.3	8.6	11.5	15.8	17.9	15.5
	NAL-NL1 target*	11.8	16.2	19.0	19.9	21.1	22.7	25.8
	Difference – 2 cc and NAL-NL1 targets	− 12.2	− 10.9	− 10.4	− 8.4	− 5.3	− 4.8	− 10.3

Abbreviations: RECD, real ear to coupler difference; REIG, real ear insertion gain; REUG, real ear unaided gain. *From Table 41.1.

Table 41.4 Calculations of the individualized coupler response for flat insertion gain (CORFIG) adjusted to include occluded venting, a 3 mm Libby horn with 1500 Ω damper, and the resulting difference between the adjusted 2 cc coupler and NAL-NL1 targets.

		Frequency (Hz)						
		250	500	1000	2000	3000	4000	6000
A. Modified individual CORFIG	Individual CORFIG*	− 3.0	− 1.1	0.4	− 0.6	2.4	3.3	2.6
	Predicted 1 mm vent	6.0	1.0	0.0	1.0	1.0	0.0	1.0
	Predicted 3 mm Libby horn with 1500 Ω damper	− 1.0	− 2.0	− 2.0	0.0	6.0	8.0	2.0
	Modified individual CORFIG	2.0	− 2.1	− 1.6	0.4	9.4	11.3	5.6
B. Comparing targets	Measured REIG**	2.6	6.4	8.2	12.1	13.4	14.6	12.9
	Modified individual CORFIG	2.0	− 2.1	− 1.6	0.4	9.4	11.3	5.6
	Adjusted 2 cc target	4.6	4.3	6.6	12.5	22.8	25.9	18.5
	NAL-NL1 target**	11.8	16.2	19.0	19.9	21.1	22.7	25.8
	Difference	− 7.2	− 11.9	− 12.4	− 7.4	1.7	3.2	− 7.3

*From Table 41.3. **From Table 41.1

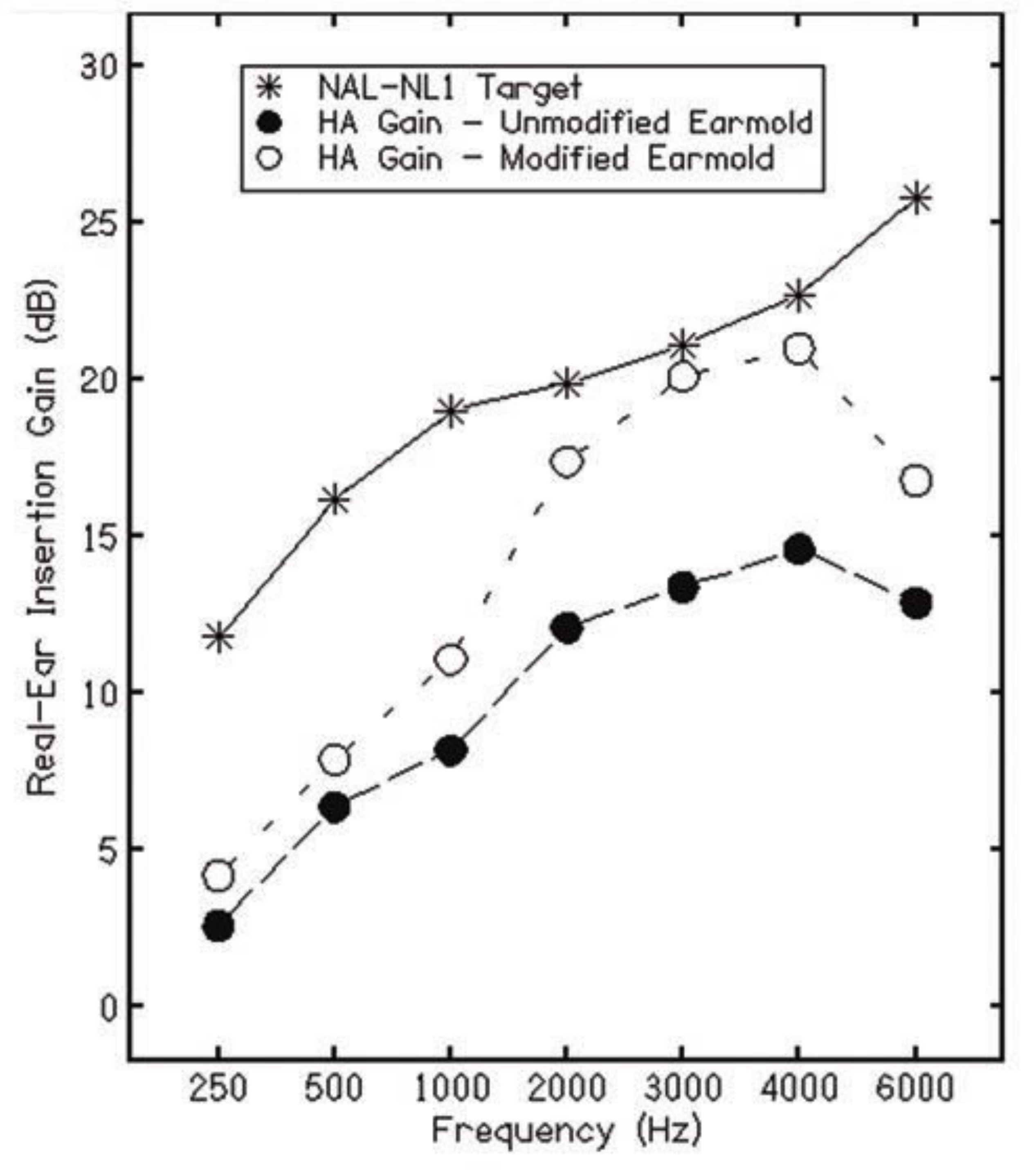


Fig. 41.2 Real ear insertion gain (REIG), in dB, representing prescriptive target for a 65 dB sound pressure level (SPL) input level (asterisk), and the REIG response of the hearing aid prior to modifications (filled circle) and after modifications (open circle).

(filled circle), and modified earmold (open circle) responses. Note that the REIG for the modified earmold better approximates target gain in the high frequencies compared with the unmodified target gain, with minimal changes in the low frequencies. At the conclusion of the session, AS reports that the hearing devices provided a marked increase in her ability to understand speech. The clinician advised AS that the modifications made were temporary, and she should meet with her audiologist for additional fine tuning when she returns home.

3. How did the clinician's actions impact the practice?
   The most important outcome from the clinician's use of his skill was his ability to reduce the communication problems experienced by AS. This created an increased level of AS's satisfaction with the clinician's help and, more importantly, with the profession. In addition, there is an increased likelihood that AS will speak highly of the service she received from this practice to her local peers.

   A secondary outcome is the increase in revenue generated by being able to provide services for the patient to result in a better outcome. Because the clinician decided to assist AS beyond the listening check, he was able to generate revenue that would not have been generated otherwise. This revenue generated can stem from either charging a fixed fee (e.g., $100) or by charging an hourly rate for professional services.

   One method to calculate the hourly rate for professional fees is based on the following equation:

$$\text{Hourly rate} = \frac{\text{Annual expenses}}{\text{Annual contact hours}} \tag{41.1}$$

   Annual expenses consist of a practice's personnel expenses (e.g., salaries, benefits) combined with clinic expenses (i.e., overhead costs, such as office supplies, rent, utilities, etc.). Annual expenses exclude expenses related to cost of goods dispensed (e.g., hearing aids, hearing assistive devices, batteries, etc.). In this case study, the annual expenses for this hypothetical clinic are $750,000.

   Annual contact hours are determined by multiplying patient contact hours/week by the number of hours worked/year times the number of providers. As an example, assume that a practice has three audiologists, working 48 weeks/year, and averaging 35 hours/week of patient contact hours. Using these data, the annual contact hours for this clinic equate to 5,040 hours (i.e., 35 hours/week × 48 weeks/year × 3 audiologists). Thus the hourly rate for this clinic equates to $148.81 (i.e., $750,000/5040 hours).

41.4 Key Points

1. Audiologists have the ability to acoustically modify the gain/output of hearing aids, to a certain degree, without the use of computer-based software. These modifications require a general knowledge about venting, dampers, and tubing length and diameter.
2. Prior to the acoustic modification of earmolds, it is imperative that the clinician also account for the contribution of the listener's individual ear canal. Failing to account for the ear canal may result in over- or underamplification and decreased patient satisfaction.
3. The example used in this case study used custom earmolds. The majority of hearing devices dispensed today, however, are receiver-in-the-canal/receiver-in-the-ear (RIC/RITE). Although the calculations may not apply directly to RIC/RITE hearing devices, the basic tenets of the tubing's internal diameter and degree of tulip dome venting still apply to gain/output modifications.

Suggested Reading

American National Standards Institute. Specification of Hearing Aid Characteristics (ANSI S3.22–2009). 2009

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42 Real-Ear Aided Response and Audibility

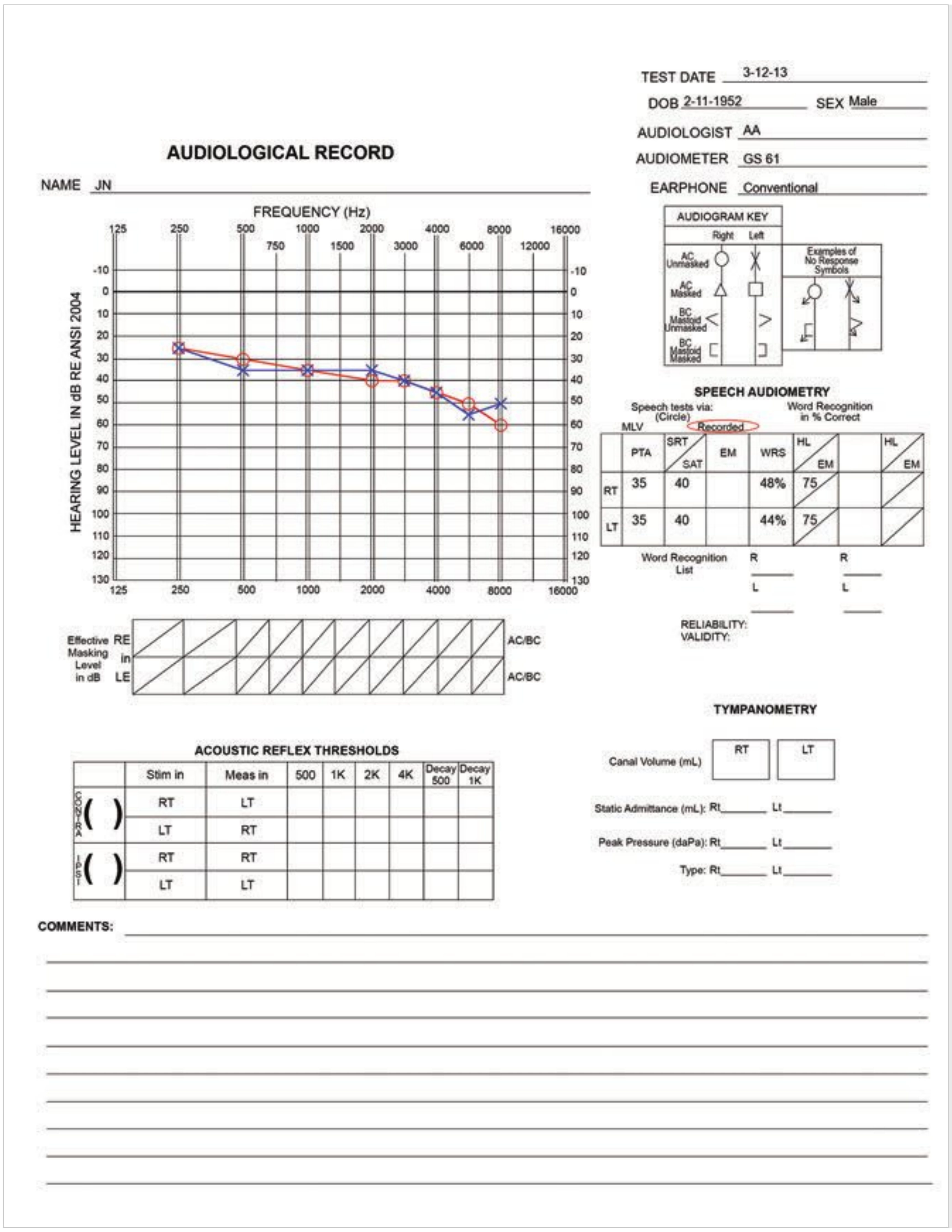
Amyn M. Amlani

This case report demonstrates the importance of using measured, not predicted, values for fitting hearing aids.

42.1 Clinical History and Description

JN is a 61-year-old tenured professor. His responsibilities include, but are not limited to, research, teaching two courses per semester, serving as an undergraduate adviser to a cohort of nearly 50 students, and providing service to the university and its local community. JN has become increasingly aware that he struggles to recognize speech on the telephone, in the classroom, and during group conversations. JN scheduled an appointment for an audiological examination at the university's speech and hearing center. Fig. 42.1 reports the results of JN's audiological examination. These results reveal a bilateral symmetrical mild to moderately severe hearing loss. Speech recognition thresholds (SRTs) revealed a mild loss in the ability to receive speech bilaterally. Word recognition scores (WRSs) revealed a very poor ability to recognize speech bilaterally. During the counseling component of the audiological examination, JN decided to pursue bilateral receiver in-the-canal (RIC) hearing aids.

The protocol at the clinic dictates that clinicians obtain individual measures of real ear to coupler difference (RECD) on all patients pursuing hearing aids prior to ordering the hearing aids. RECD is the measured difference as a function of frequency in sound pressure level (SPL) between the 2 cc coupler, which is a small-volume cavity used as a quality control means for hearing aid electroacoustic measurement, and the patient's ear canal. The 2 cc coupler measure is performed in a test box and does not require the presence of the patient. Conversely, the real-ear component of the RECD requires the careful



AUDIOLOGICAL RECORD

NAME JN

TEST DATE 3-12-13

DOB 2-11-1952 SEX Male

AUDIOLOGIST AA

AUDIOMETER GS 61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	35	40		48%	75		
LT	35	40		44%	75		

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt____ Lt____
Peak Pressure (daPa): Rt____ Lt____
Type: Rt____ Lt____

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 42.1 Audiological results for JN.

placement of a probe tube in the patient's ear canal. In adult patients, the 2 cc coupler, with its smaller volume than the average adult ear canal and its inability to duplicate individual differences in acoustic impedance of the human ear canal, will typically underestimate the SPL measured in the average human ear canal. Even with these known differences, the clinician is "running behind" and cannot obtain individual measures.

Once the hearing aids arrive, it is the clinician's responsibility to verify that the hearing aids match the manufacturer's specifications and to program the hearing aids for the subsequent fitting. In this case report, the hearing aids' electroacoustic performance is verified to match the manufacturer's specifications and Memory 1 in the hearing aids is programmed to match National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1). During initial programming, the additional memories and features are disabled. JN returned for his initial hearing aid fitting, at which time the devices are fit and verified using probe-microphone measures. Fig. 42.2 reports the audiometric thresholds for the right ear, converted to dB SPL (asterisks) as well as the prescribed real ear aided response (REAR) for NAL-NL1 for an input level of 65 dB SPL for a bilateral fit using a three-channel hearing aid (filled stars). Also reported is the measured REAR using the predicted RECD (filled circle). At the fitting, JN stated he was disappointed that his new devices did not appreciably improve his recognition of speech in everyday listening environments.

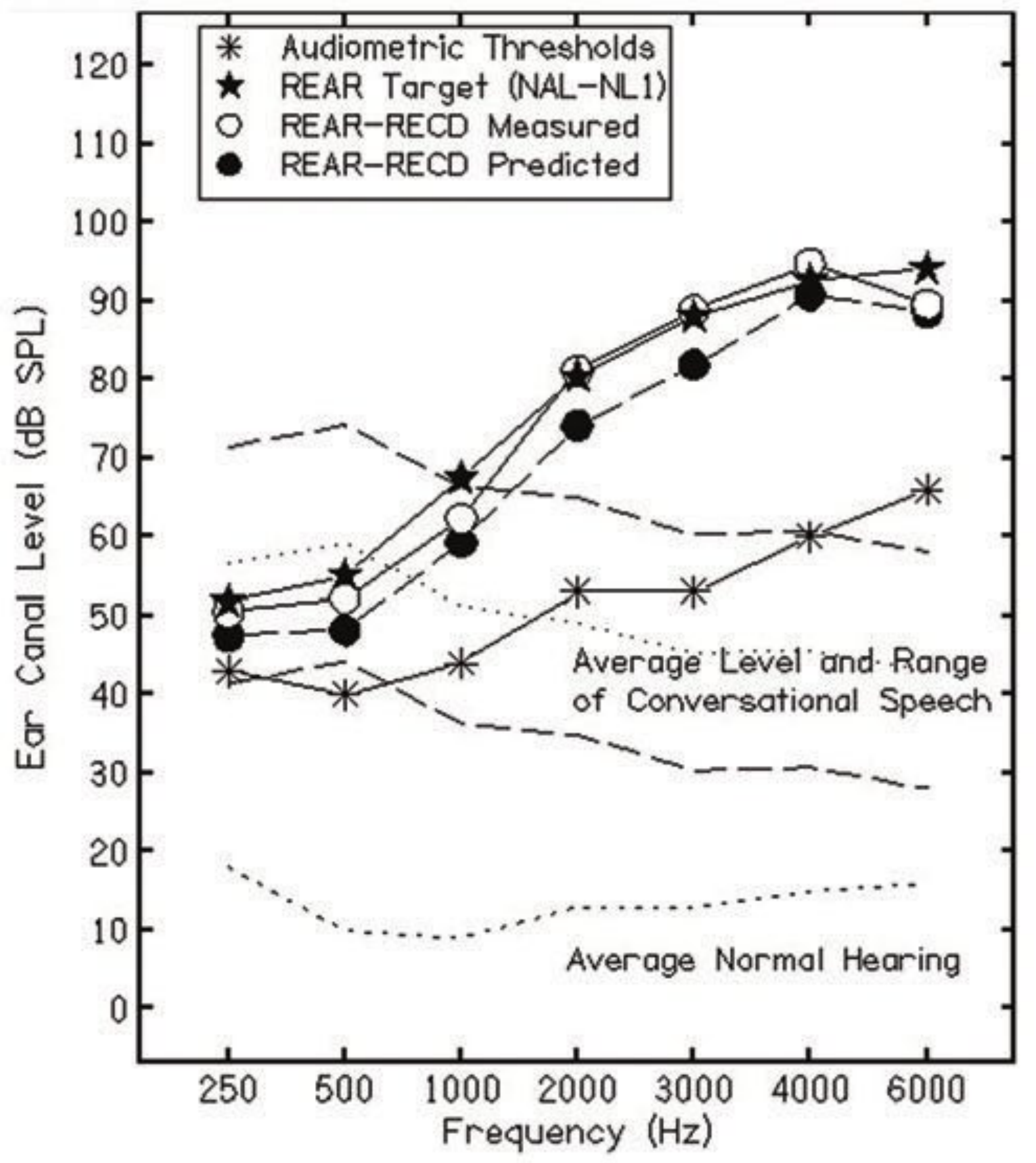


Fig. 42.2 Audiogram, in dB sound pressure level (SPL), representing JN's audiometric thresholds for the right ear (asterisk), the NAL-NL1 real ear aided response (REAR) prescriptive target for a 65 dB SPL input level (star), and the REAR of the hearing aid based on measured real ear to coupler difference (RECD) (open circle), and predicted RECD (filled circle) values.

Table 42.1 Predicted and individually measured real ear to coupler differences (RECDs)

	Frequency (Hz)						
	250	500	1000	2000	3000	4000	6000
Predicted	3	4	8	7	8	13	13
Measured	6	8	11	14	15	17	14

42.2 Questions to the Reader

1. Were JN's individual RECD values similar to the predicted RECD values?
2. If the RECD values are different, how do the predicted and individual measures influence the hearing aid fitting as measured by REAR?
3. To what degree do the predicted and individual measures of RECD influence audibility?
4. How does JN's initial response to amplification influence hearing aid adoption?

42.3 Discussion of Questions to the Reader

1. Were JN's individual RECD values similar to the predicted RECD values?

   Table 42.1 reports predicted RECD and individually measured RECD. The differences between these RECD values

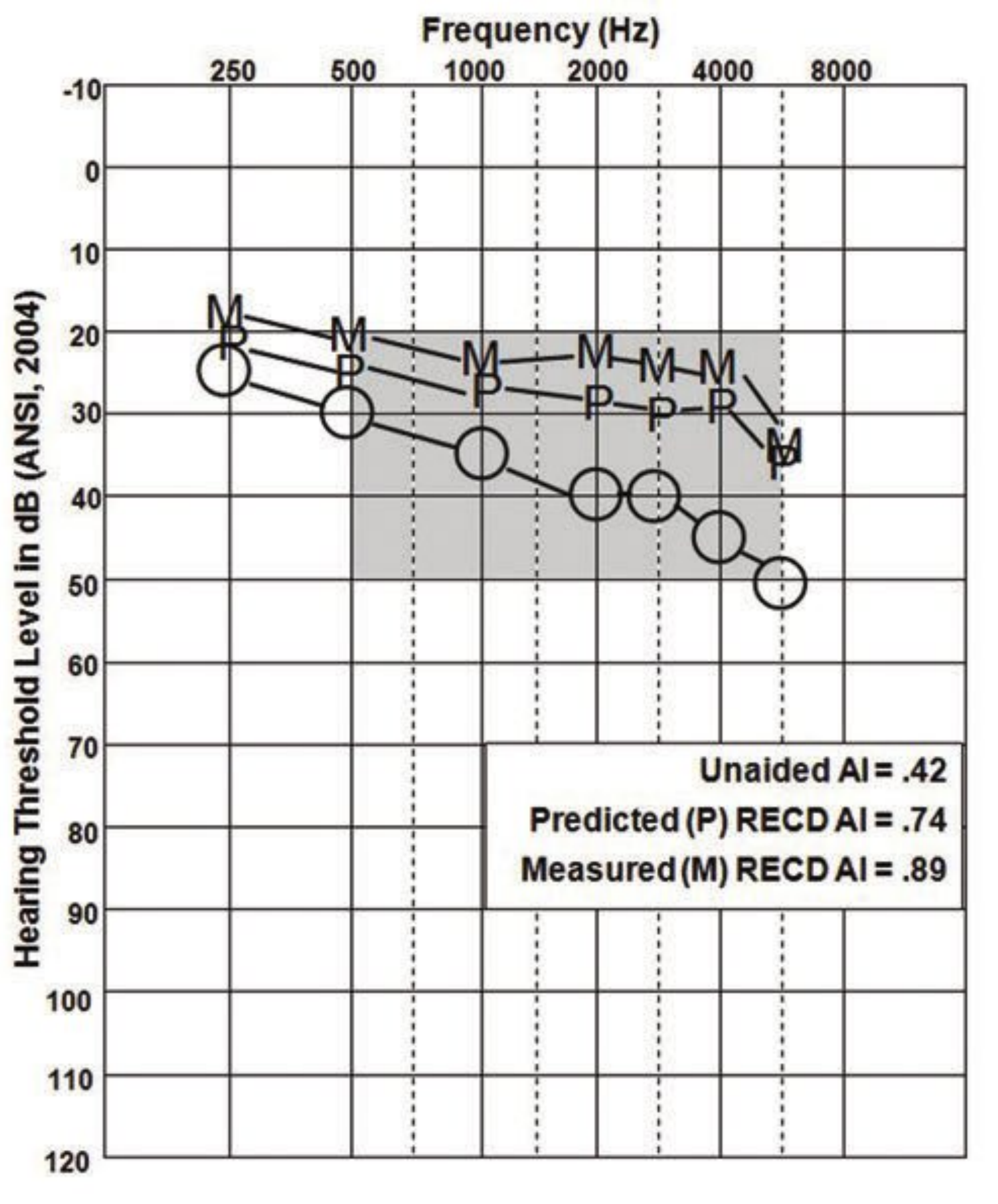


Fig. 42.3 Audibility calculations, based on Pavlovic's (1988) $A_0(6)$ procedure, for unaided (circle), predicted (P) real ear to coupler difference (RECD), and measured (M) RECD.

indicate that JN's ear canal response deviated markedly from the predictive ear canal response. Specifically, JN's RECD values are markedly greater than the predicted values at all test frequencies except for 6000 Hz, indicating that the SPL produced in JN's ear canal is significantly greater than the SPL measured in a 2 cc coupler. The larger measured RECD values found for JN indicate that JN's ear canal volume is significantly smaller than the volume of the 2 cc coupler used to measure hearing aid performance.

2. If the RECD values are different, how do the predicted and individual measures influence the hearing aid fitting as measured by REAR?
   Because the clinician could not account for the individual RECD, the hearing aid in JN's case was programmed with insufficient gain/output. As reported in Fig. 42.2, the REAR response of the hearing aid using predicted RECD (filled circle) is markedly lower than the REAR response using measured RECD (open circles), and the differences in magnitude is equal to the differences between predicted and measured RECD in Table 42.1. Had the clinician obtained individual RECD measures, the initial REAR would have better approximated the REAR NAL-NL1 target (filled stars) in Fig. 42.2.
3. To what degree do the predicted and individual measures of RECD influence audibility?

Fig. 42.3 illustrates the influence of predicted and individual RECD measures on audibility for JN. In this case report, audibility is quantified using the $A_o(6)$ method originally described by Pavlovic. Audibility is quantified on a scale from 0.00 to 1.00, with values closer to 0.00 representing no availability of speech cues and 1.00 representing the availability of all speech cues to the listener. Audibility is sometimes shown as a percentage and derived by multiplying the audibility value by 100 (e.g., audibility = 0.38 or 38% [0.38 × 100]).

The $A_o(6)$ method quantifies audibility using an audiogram, under the premise that speech cues important to speech recognition range between 20 and 50 dB (i.e., 30 dB dynamic range of speech) at 250 to 6000 Hz. This region is shaded in Fig. 42.3. To illustrate the use of this clinical tool, the unaided threshold (O-O) in Fig. 42.3 at 500 Hz is 30 dB HL. At this frequency, only 20 dB of audibility (i.e., 50 dB HL − 30 dB HL) is available to JN. At 1000, 2000, 3000, 4000, and 6000 Hz, audibility is 15 dB, 10 dB, 10 dB, 5 dB, and 0 dB, respectively. The amount of speech available to JN for unaided listening is 0.42 (lower box to the right). The amount of audibility available to JN in the unaided condition was derived by summing the amount of audibility at the three lower frequencies (i.e., 20 + 15 + 10 = 45) and adding this to

Table 42.2 Real ear aided response (REAR) calculations, and the corresponding audibility, using predicted real ear to coupler difference (RECD) values

		Frequency (Hz)						
		250	500	1000	2000	3000	4000	6000
A. Convert dB HL to dB SPL	Thresholds (dB HL)	25	30	35	40	40	45	50
	MAP corrections	+18	+10	+9	+13	+13	+15	+16
	Thresholds (dB SPL)	43	40	44	53	53	60	66
B. Calculate REAR – Predicted (P)	Thresholds (dB SPL)	43	40	44	53	53	60	66
	RECD-predicted (P)	+3	+4	+8	+7	+8	+13	+13
	Mic effect (BTE)	+0.5	+1.2	+0.3	+4.1	+2.8	+3.7	+1.6
	REUG	+1	+3	+7	+10	+18	+14	+8
	REAR – Predicted (P)	47.5	48.2	59.3	74.1	81.8	90.7	88.6
C. Calculate REIG from REAR	REAR – Predicted (P)	47.5	48.2	59.3	74.1	81.8	90.7	88.6
	Thresholds (dB SPL)	− 43	− 40	− 44	− 53	− 53	− 60	− 66
	REUG	− 1	− 3	− 7	− 10	− 18	− 14	− 8
	REIG	3.5	5.2	8.3	11.1	10.8	16.7	14.6
D. Calculate aided threshold	Thresholds (dB HL)	25	30	35	40	40	45	50
	REIG	− 3.5	− 5.2	− 8.3	− 11.1	− 10.8	− 16.7	− 14.6
	Aided threshold	21.5	24.8	26.7	28.9	29.2	28.3	35.4
E. Audibility calculation	Aided threshold	21.5	24.8	26.7	28.9	29.2	28.3	35.4
	Audibility	28.5	25.2	23.2	21.1	20.8	21.7	14.6
	$A_o(6)$ method	0.89						

Abbreviations: BTE, behind-the-ear; HL, hearing level; MAP, Minimal Audible Pressure; REAR, real ear aided response; RECD, real ear to coupler difference; REIG, real ear insertion gain; SPL, sound pressure level.

the average audibility in the higher frequencies (i.e., [10 + 5 + 0]/3 = 5). Together, these values equal 50 (i.e., 45 + 5), which is divided by 120, or the maximum number of audible decibels.

In Fig. 42.3 note that audibility has also been calculated (box to the lower right) for the predicted and measured RECD aided conditions, which are denoted as P and M, respectively. The aided audibility values were derived using the same $A_o(6)$ method already described, with data derived by transforming REAR data to real ear aided gain (REAG; derived as REAR minus input signal [obtained at 65 dB in this example]) and subtracting the patient's real ear unaided gain (REUG). These transformations result in real ear insertion gain (i.e., REIG = REAG − REUG), which is then subtracted, when REIG is positive, from unaided thresholds.

For aided audibility, results indicate that the hearing aid using the predicted RECD provided JN with 74%(i.e., AI = 0.74) of the available speech information, or an improvement of 0.32 compared with the unaided condition. Had the clinician used the individually measured RECD, the aided audibility would have improved to 0.89, or a 0.47 increase in the availability of speech sounds compared with the unaided condition. Calculations for the predicted and measured RECD aided conditions are reported in Table 42.2 and Table 42.3, respectively.

4. How does JN's initial response to amplification influence hearing aid adoption?

A recent study revealed hearing aid adoption is dependent on perceived value. Perceived value is defined as the patient's assessment of the utility of the product and is dependent on what the product provides to the end user relative to the price paid. If perceived value is high, then the likelihood of adoption increases. Conversely, if perceived value is low, then the likelihood of adoption decreases.

Given that JN's initial reaction to amplification, programmed to predicted RECD values, was disappointment, it can be assumed that perceived value decreased and most likely reduced the likelihood of hearing aid adoption. If the clinician had measured individual RECD and implemented

Table 42.3 Real ear aided response (REAR) calculations and the corresponding audibility, using measured real ear to coupler difference (RECD) values

		Frequency (Hz)						
		250	500	1000	2000	3000	4000	6000
A. Convert dB HL to dB SPL	Thresholds (dB HL)	25	30	35	40	40	45	50
	MAP corrections	+18	+10	+9	+13	+13	+15	+16
	Thresholds (dB SPL)	43	40	44	53	53	60	66
B. Calculate REAR – Measured (M)	Thresholds (dB SPL)	43	40	44	53	53	60	66
	RECD − Measured (M)	+6	+8	+11	+14	+15	+17	+14
	Mic effect (BTE)	+0.5	+1.2	+0.3	+4.1	+2.8	+3.7	+1.6
	REUG	+1	+3	+7	+10	+18	+14	+8
	REAR – Measured (M)	50.5	52.2	62.3	81.1	88.8	94.7	89.6
C. Calculate REIG from REAR	REAR – Measured (M)	50.5	52.2	62.3	81.1	88.8	94.7	89.6
	Thresholds (dB SPL)	− 43	− 40	− 44	− 53	− 53	− 60	− 66
	REUG	− 1	− 3	− 7	− 10	− 18	− 14	− 8
	REIG	6.5	9.2	11.3	18.1	17.8	20.7	15.6
D. Calculate aided threshold	Thresholds (dB HL)	25	30	35	40	40	45	50
	REIG	− 6.5	− 9.2	− 11.3	− 18.1	− 17.8	− 20.7	− 15.6
	Aided threshold	18.5	20.8	23.7	21.9	22.2	24.3	34.4
E. Audibility calculation	Aided threshold	18.5	20.8	23.7	21.9	22.2	24.3	34.4
	Audibility	30	29.2	26.3	28.1	27.8	25.7	15.6
	$A_o(6)$ method	0.89						

Abbreviations: Abbreviations: BTE, behind-the-ear; HL, hearing level; MAP, Minimal Audible Pressure; REAR, real ear aided response; RECD, real ear to coupler difference; REIG, real ear insertion gain; SPL, sound pressure level.

those measures during the fitting process, JN's initial reaction to amplification probably would have enhanced JN's satisfaction. The increase in satisfaction, in turn, would probably have increased the perceived value and, ultimately, hearing aid adoption. In addition to hearing aid adoption, the clinician inadvertently may have lost the opportunity of JN referring other patients and, possibly, hearing aid users.

42.4 Key Points

1. Successful hearing aid fittings are dependent on accounting for individual differences between 2 cc coupler measures and the deviations inherent in the human ear canal. Failure to account for these differences may result in an unsatisfactory trial period with hearing aids, which may reduce the likelihood of hearing aid adoption.
2. Hearing aid adoption, patient satisfaction, and reduced patient visits can be enhanced by obtaining individual measures of RECD, which, when implemented to the REAR prescriptive target, can improve audibility. Hearing aid adoption and patient satisfaction are attributes known to inspire patients to refer family members and friends for audiological treatment.

Suggested Reading

Amlani AM. Influence of perceived value on hearing aid adoption and re-adoption intent. Hearing Products Review 2013;(March 8–12)

Amlani AM, Punch JL, Ching TC. Methods and applications of the audibility index in hearing aid selection and fitting. Trends Amplif 2002; 6: 81–130

Bagatto MP, Moodie S, Scollie SD et al. Clinical protocols for hearing instrument fitting in the desired sensation level method. Trends Amplif 2005; 9: 199–226

Byrne D, Dillon H, Ching T, Katsch R, Keidser G. NAL-NL1 procedure for fitting nonlinear hearing aids: characteristics and comparisons with other procedures. J Am Acad Audiol 2001; 12: 37–51

Pavlovic CV. Articulation index predictions of speech intelligibility in hearing aid selection. ASHA 1988; 30: 63–65

E-GIRQ.COM

43 Medical Professional with Hearing Loss Seeking Amplified Stethoscope

A. U. Bankaitis and Pamela A. Ison

DG is a 58-year-old female nurse practitioner seeking a stethoscope that will amplify breath/lung and heart sounds to compensate for her hearing loss.

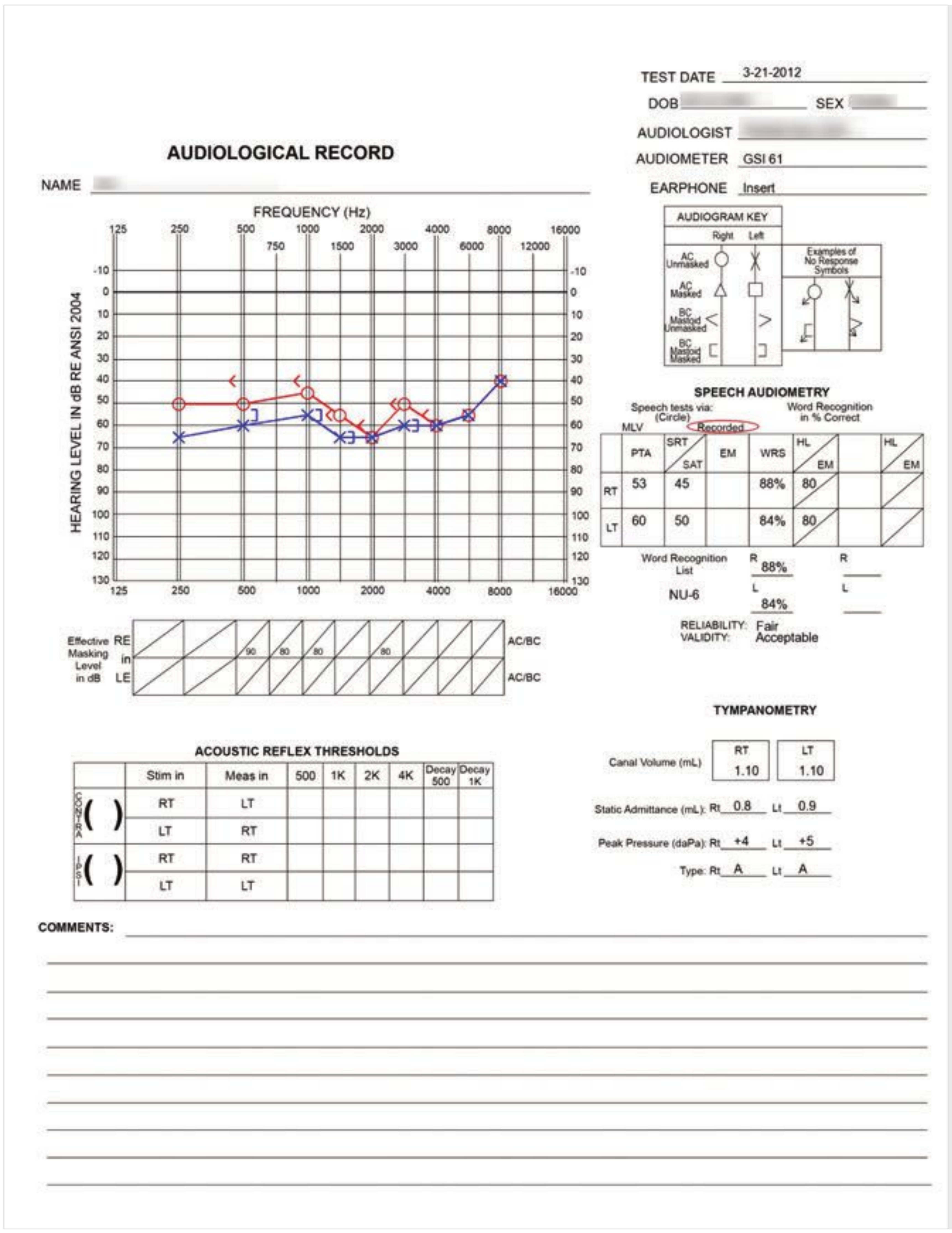
43.1 Clinical History and Description

DG is a 58-year-old nurse practitioner with a long-standing history of bilateral sensorineural hearing loss and vertigo. DG's hearing loss was initially detected 7 years ago at another facility, at which time she was diagnosed with Ménière disease. Her hearing loss has reportedly remained stable over time; however, DG recently experienced several episodes of vertigo along with a perceived decrease in her hearing, which precipitated an otology consult.

With her vertigo under control, DG was medically cleared and referred to an audiologist for management of her hearing loss. DG was primarily concerned about her inability to detect breath/lung and heart sounds during standard auscultation procedures. She was interested in exploring amplified stethoscope options to assist in performing routine clinical procedures inherent to daily job responsibilities.

43.2 Audiological Testing

The audiological examination (Fig. 43.1) revealed a bilateral symmetrical moderate to moderately severe sensorineural hearing loss that is flat in configuration. Speech recognition thresholds (SRTs) were consistent with pure-tone findings bilaterally and revealed a moderate loss in the ability to receive



AUDIOLOGICAL RECORD

NAME [redacted]

TEST DATE 3-21-2012

DOB [redacted] SEX [redacted]

AUDIOLOGIST [redacted]

AUDIOMETER GSI 61

EARPHONE Insert

Effective Masking Level in dB	RE in			90	80	80		80			AC/BC
	LE										AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	53	45		88%	80		
LT	60	50		84%	80		

Word Recognition List NU-6

R 88% R

L 84% L

RELIABILITY: Fair

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.10	1.10

Static Admittance (mL): Rt 0.8 Lt 0.9

Peak Pressure (daPa): Rt +4 Lt +5

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 43.1 Audiometric thresholds (dB HL) of DG obtained immediately prior to initial consultation.

speech. Word recognition scores (WRSs) were obtained bilaterally at a presentation level of 30 dB sensation level (SL) with scores of 88% and 84% for the right and left ears, respectively. These WRSs indicate a slight loss in the ability to recognize speech bilaterally. Immittance audiometry indicated normal tympanic membrane mobility bilaterally as indicated by type A tympanograms. Ear canal volume (mL), static compliance (mL), and middle ear pressure (daPa) were within normal limits bilaterally. Contralateral and ipsilateral acoustic reflex thresholds (ARTs) testing and reflex decay were not performed.

43.3 Questions to the Reader

1. What viable amplified stethoscope options are available to a patient who does not wear a hearing instrument and has DG's magnitude and configuration of hearing loss?
2. What additional counseling is needed prior to proceeding with a recommendation for an amplified stethoscope?
3. What potential amplified stethoscope options are available with receiver-in-the-ear (RITE) hearing instruments?
4. What hearing instrument programming adjustments should be considered when using an amplified stethoscope in combination with hearing instruments?

43.4 Discussion of Questions to the Reader

1. What viable amplified stethoscope options are available to a patient who does not wear a hearing instrument and has DG's magnitude and configuration of hearing loss?
   Because DG is not currently using hearing instrumentation, any one of several commercially available stand-alone amplified stethoscopes (i.e. 3 M Littmann, 3 M, St. Paul, MN; Adscope 657, American Diagnostic, Hauppauge, NY; E-Scope II, Cardionics, Webster, TX) may be recommended. Equipped with traditional earpieces, stand-alone amplified stethoscopes are used in the same fashion as traditional stethoscopes. The only operational difference between an amplified and traditional stethoscope is that amplified stethoscopes are battery-operated devices because they must amplify breath/lung and heart, or other body sounds to compensate for as much as a severe hearing loss. Which stand-alone stethoscope to recommend remains a matter of patient preference, which may be influenced by factors including, but not limited to product price, available features that appeal to the user (i.e., ability to record body sounds for later reference), and product warranty. Table 43.1 lists the features

Table 43.1 Features of commercially available stand-alone amplified stethoscopes

Device	Features
 3 M Littman Amplified Electronic Stethoscope (3 M Corporation, St. Paul, MN)	• 2-year warranty • 25 dB of gain • Can record/playback auscultation • Transmits recordings wireless to other comparable 3 M Littman stethoscopes • Approximate MSRP of $575.00
 E-Scope II Amplified Stethoscopes with Earpieces (Cardionics, Inc., Webster, TX)	• 1-year warranty • 50 dB of gain with 130 dB sound pressure level max output • Modified versions available to accommodate hearing instrument use • Approximate MSRP of $335.00
 Adscope 657 Electronic Stethoscope (American Diagnostic Corp., Hauppauge, NY)	• 2-year warranty • 24 dB of gain • Approximate MSRP of $260

of three currently available stand-alone amplified stethoscopes.

2. What additional counseling is needed prior to proceeding with a recommendation for an amplified stethoscope?
   DG was primarily interested in pursuing an amplified stethoscope to compensate for her hearing loss as it related to measuring her patients' blood pressure. Despite a longstanding history of hearing loss, DG exhibited little, if any, regard to her daily communication needs beyond performing a specific job function. The degree of her hearing loss presented significant communication challenges both at and beyond the work environment. Rather than immediately commence with an orientation to potential amplified stethoscope solutions it was necessary for the audiologist to counsel DG about her need for and the associated benefits of bilateral amplification. Once the primary amplification needs were outlined and addressed, DG's secondary communication needs associated with identifying viable solutions for performing auscultation procedures were addressed. Hearing instrument options were reassessed as amplified stethoscopes were discussed to find the best solution that would meet DG's communication needs at home and work. The decision was eventually made to proceed with bilateral RITE hearing instruments and to purchase an amplified stethoscope specifically designed to integrate with these hearing instruments.
3. What potential amplified stethoscope options are available with receiver-in-the-ear (RITE) hearing instruments?
   The most straightforward option involves removal of the hearing instruments prior to auscultation and relying on the use of a stand-alone amplified stethoscope. One drawback of this approach is the need to remove and reinsert hearing instruments prior to and then following auscultation procedures. A second option is to use a stand-alone amplified stethoscope with traditional earpieces while continuing to wear the hearing instruments. Because RITE instruments can leave the ear canal unoccluded, and depending on ear canal dimensions and an individual's comfort tolerances, it may be possible to use a stand-alone amplified stethoscope without having to remove the hearing instruments. The third option is to purchase an amplified stethoscope that is modified to interface with hearing instruments.

   Currently, there are two commercially available amplified stethoscopes that can be retrodesigned to interface with hearing instruments through the use of additional accessories or via Bluetooth technology or both. First, the E-Scope II (Model 718–7710) is a modified version of the stand-alone model whereby the traditional earpieces have been removed and replaced with a pair of headphones that are hard-wired to the stethoscope amplifier and then worn over the ears (Fig. 43.2). This stethoscope can provide 50 dB of gain with a maximum output of 130 dB sound pressure level (SPL). Second, the recently available Audiologist's Choice Bluetooth-amplified stethoscope (AC-Scope) (Fig. 43.3) is designed to specifically work with hearing instruments communicating with a Bluetooth streamer. This stethoscope can provide 24 dB of gain. Once the stethoscope and streamer are paired, breath/lung and heart sounds are wirelessly transmitted from the stethoscope's amplifier directly to the user's streamer. The streamer, in turn, delivers body sounds directly to the hearing instruments.

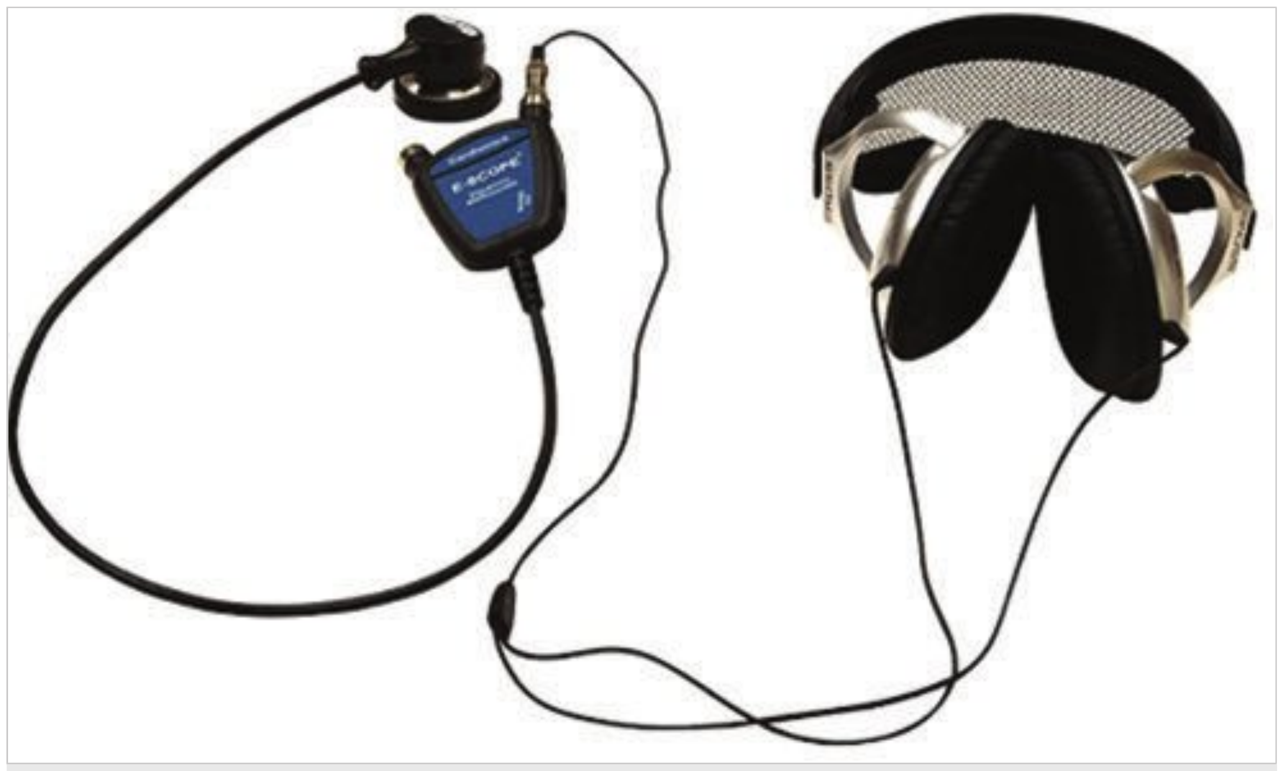


Fig. 43.2 Modified E-Scope II (Model 718–7712) with oversized headphones manufactured by Cardionics (www.Cardionics.com). (Used with permission from Cardionics.)

4. What hearing instrument programming adjustments should be considered when using an amplified stethoscope in combination with hearing instruments?
   Although there are some differences of opinion regarding the frequency range of various body sounds, the frequency range of breath/lung sounds is from 70 to 4000 Hz, with the most critical sounds used for differential diagnosis at 200 to 600 Hz. The frequency components of heart sounds are lower, typically at 20 to 650 Hz, although 70 to 120 Hz represents the most critical frequency range of heart sounds for the healthcare professional to hear. Although an audiometric configuration may dictate amplifying the high-frequency region for optimizing speech, audiologists need to consider providing a low-frequency emphasis program for healthcare professionals with hearing loss because both breath/lung and heart sounds have low-frequency emphasis. Although specific data may be lacking, it is accurate to report that breath/lung and heart sounds represent extremely soft sounds. Ideally, a hearing instrument capable of having low compression

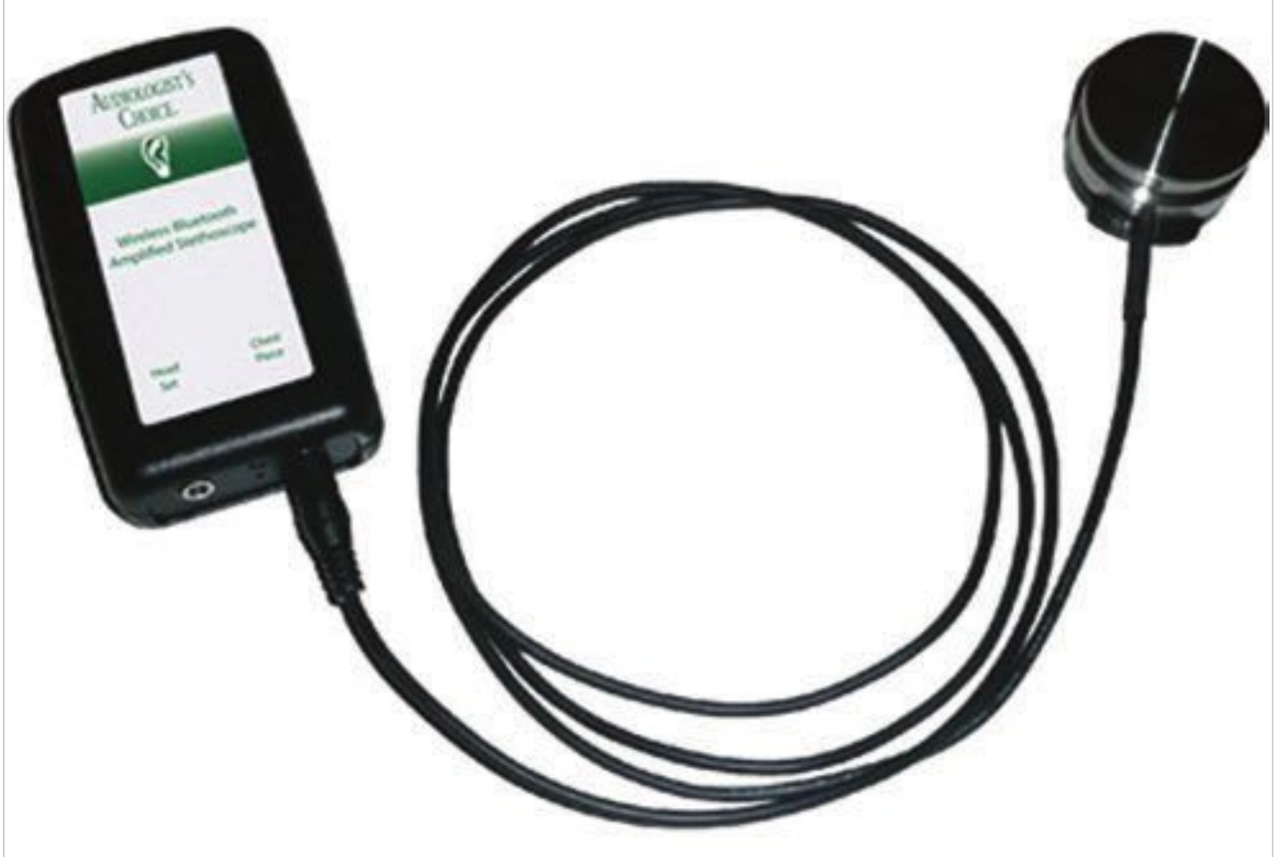


Fig. 43.3 The Audiologist's Choice Bluetooth amplified stethoscope (AC-Scope) available through Oaktree Products, Inc. (www.oaktree-products.com). (Used with permission from Oaktree Products, Inc.)

thresholds using wide dynamic range compression (WDRC) would be beneficial because this method of compression provides greater gains/output for soft input sounds.

43.5 Diagnosis and Recommended Treatment

Although DG was motivated to pursue an amplified stethoscope, she had not considered the degree to which her hearing loss impacted her life beyond the ability to perform a specific job function. At the time of the initial consultation, communication needs were prioritized and outlined with the recommendation to pursue bilateral amplification. Once DG accepted the need to pursue bilateral amplification, hearing instrument technology options were addressed, taking into consideration potential amplified stethoscope solutions associated with each hearing instrument option.

DG was fit bilaterally with Intiga 8 RITE hearing instruments (Oticon, Somerset, NJ) equipped with a ConnectLine streamer to specifically accommodate her preference to pursue the AC-Scope. DG was counseled that, as a recently available device, the AC-Scope had been successfully paired with a handful of hearing instrument streamers and the stability of the wireless transmission during typical clinical applications remained unknown. For example, some users could not successfully pair the AC-Scope to their streamer. Others reported that the Bluetooth signal transmission from the stethoscope’s amplifier to their streamer was intermittent, precluding successful use of the device in a wireless manner. If either was the case, DG was made aware that the Bluetooth capabilities of the AC-Scope could be bypassed by directly connecting the stethoscope amplifier to the streamer via an audio cord. The AC-Scope amplifier contains an additional headphone to enable the audiologist to simultaneously listen to breath/lung and heart sounds along with the healthcare professional to optimize hearing instrument programming. In the event of Bluetooth transmission issues, the headphone port of the AC-Scope may be used to connect the stethoscope amplifier directly to the streamer with a special audio-in cable packaged with the AC-Scope. It is important to add that an additional audio cord accessory available from the hearing instrument manufacturer may also be necessary to properly interface the AC-Scope to the streamer.

43.6 Outcome

DG reported substantial benefit from her hearing instruments, including the realization of the extent that her hearing loss, when unaided, exhausted her. In addition, DG was able to successfully use the AC-Scope wirelessly with her hearing instruments during auscultation procedures. Unfortunately, the device occasionally lost connectivity with the streamer, and it was necessary to hard-wire the stethoscope to the streamer. Despite this hard-wired connection, the configuration was still plagued with intermitted signal transmission for reasons that remained unknown and unresolved. As a result, DG was subsequently fit with a modified version of the E-Scope II equipped with oversized headphones, a solution that met her needs. Interestingly, the patient initially decided to keep both stethoscopes, preferring and primarily using the AC-Scope and switching to the E-Scope II in the presence of a connection failure. Eventually, DG decided to use only the E-Scope II.

43.7 Key Points

Recently, the Audiologist s Choice Bluetooth Amplified Stethoscope (AC-Scope) was voluntarily removed from the market due to Bluetooth signal transmission issues that either precluded necessary pairing between the AC-Scope s amplifier and the hearing instrument streamer or, even when successfully paired, resulted in intermittency sufficient to interfere with performing auscultation procedures. At this time, there are no known amplified stethoscopes commercially available with Bluetooth capabilities specifically designed for pairing with a hearing instrument.

1. Counseling healthcare professionals with hearing loss on hearing instrument options and amplified stethoscopes should not be mutually exclusive events.
2. The chances of successfully meeting all of a patient’s communication needs increase when both technologies (i.e., hearing instruments and amplified stethoscopes) are considered.
3. A specific program in the hearing aids is necessary to enable detection of the very soft and low-frequency breath/lung and heart sounds associated with auscultation.
4. Identifying the best amplified stethoscope solution for healthcare professionals with hearing loss should involve access to and the ability for the patient to try two or three potential options.
5. The E-Scope II can be hard-wired to a hearing instrument streamer, eliminating the need for the patient to use headphones. It is necessary to purchase a “special” patch cord from Cardionics and to use the audio cable accessory packaged with the streamer to connect the amplifier of the stethoscope to the streamer. Unfortunately, this configuration has been associated with signal degradation that may be sufficient to interfere with successful execution of auscultation.

Suggested Reading

Abrams J. Essentials of Cardiac Physical Diagnosis. Philadelphia, PA: Lea and Febiger; 1987

Rennert, N.J, Morris, R., Barrere, C. (published March 14, 2008). How to Cope with Scopes: Stethoscope Selection and Use with Hearing Aids and CIs. Available at: http://www.hearingreview.com/practice-management/15830-how-to-cope-with-scopes-stethoscope-selection-and-use-with-hearing-aids-and-cis

44 Fitting BICROS to a Patient with Long-Standing Unilateral Hearing Loss

Alison M. Brockmeyer-Lauer

AG is a 67-year-old female with long-standing unilateral hearing loss (UHL) in the right ear, following surgery to remove a right acoustic tumor approximately 15 years ago. She has noticed a decline in hearing in her left ear and began investigating amplification options. She was fit with a Phonak Audeo S Smart III (Phonak U.S., Warrenville, IL) open-fit receiver-in-the-canal (RIC) aid in the left ear with a Phonak CROS transmitter on the right ear.

44.1 Clinical History and Description

AG is a 67-year-old female who lost all hearing in her right ear following surgery to remove a right acoustic tumor approximately 15 years ago. She reports she adjusted well to hearing only with her left ear following the surgery and functioned well without hearing assistive devices. She is active in an acoustic tumor support group and has always been comfortable with explaining her hearing loss to communication partners and asking others to sit on her left side. She reports, however, that hearing in her left ear is declining and she has been noticing greater difficulty communicating in the last year. She is now interested in hearing assistive devices to make it easier for her to communicate. She reports tinnitus in her left ear, but states she does not usually notice the tinnitus. She denies dizziness, otalgia, familial history of hearing loss, or a history of noise exposure, but she reports occasional pressure in the right ear.

44.2 Audiological Testing

Audiological examination of the left ear revealed slight sensorineural hearing loss (SNHL) at 250 Hz, rising to hearing sensitivity within normal limits from 500 to 2000 Hz, sloping from mild SNHL at 3000 and 4000 Hz to a moderate SNHL at 6000 and 8000 Hz. The right ear presented with no measurable hearing within the limits of the audiometer (Fig. 44.1). A speech recognition threshold (SRT) of 15 dB HL was obtained in the left ear, consistent with the pure-tone thresholds in the left ear. No response was obtained to spondee words during assessment of the SRT in the right ear. The word recognition score (WRS) in the left ear was obtained using a recorded version of Northwestern University Auditory Test No. 6 (NU-6) monosyllabic word lists with a female speaker. Words were presented at most intelligible level (MIL), which corresponded to 45 dB sensation level (SL) relative to the SRT. A score of 100% was obtained in the left ear, demonstrating normal ability to recognize speech. Word recognition ability could not be measured in the right ear.

Immittance audiometry was performed and tympanograms were within normal limits bilaterally. Ear canal volume (mL), static admittance (mL), and middle ear pressure (daPa) were all within the normal range bilaterally. Ipsilateral acoustic reflex thresholds (ARTs) at 500 to 4000 Hz in the left ear were within normal limits. Contralateral ARTs stimulating the left ear were present within normal limits at 1000 Hz, present at elevated levels at 500 and 2000 Hz, and absent at 4000 Hz. Ipsilateral and contralateral ARTs stimulating the right ear were absent at 500 to 4000 Hz. Contralateral acoustic reflex decay stimulating the left ear was within normal limits.

44.3 Questions to the Reader

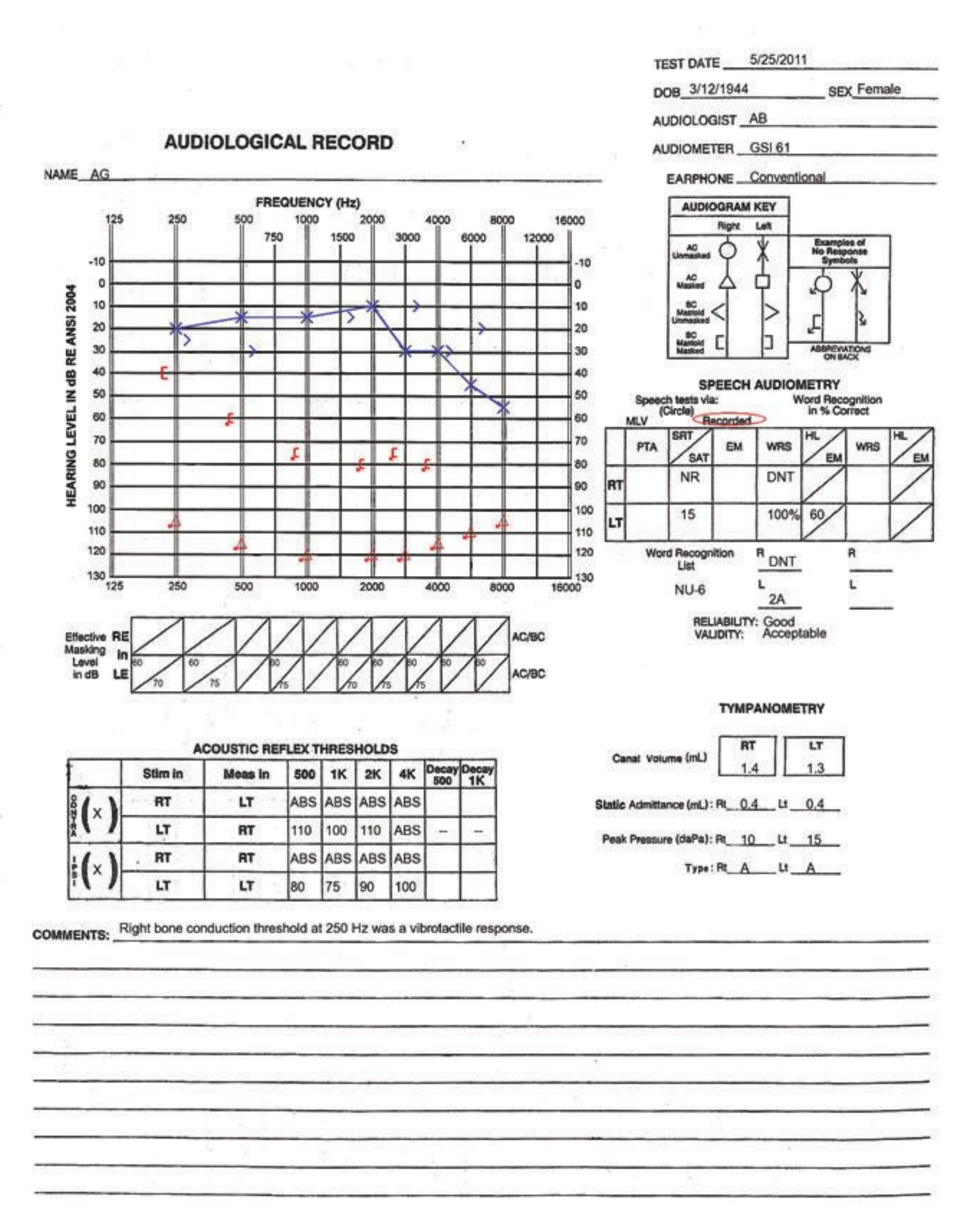
1. How does UHL affect communication? How would a mild to moderate high-frequency hearing loss in the better ear further impair communication?
2. What treatment options are available for UHL and which option would be most appropriate for this patient with a mild to moderate high-frequency hearing loss in the better ear?
3. How would a clinician counsel the patient on the realistic expectations of options for amplification for UHL?
4. What type of verification is needed to fit BICROS amplification?

44.4 Discussion of Questions to the Reader

1. How does UHL affect communication? How would a mild to moderate high-frequency hearing loss in the better ear further impair communication?

   There are several benefits of binaural hearing, including binaural loudness summation, improved recognition of speech in noisy environments, and improved localization of sound. Binaural loudness summation simply means the loudness of a sound is greater if sound is heard by two ears when compared with being heard by one ear. Studies show that binaural summation increases loudness by approximately 3 to 6 dB. Therefore, a softer sound can be more easily heard with two ears than with only one ear.

   Improved recognition of speech in noisy environments arises from head diffraction, and the central phenomena of binaural squelch and binaural redundancy. Head diffraction, or the head shadow effect, simply means that sound on one side of the head has to bend around the head in order to get to the ear on the opposite side. This will reduce the intensity of the sound reaching the opposite ear. A person can then attend to the ear with the better signal to noise ratio (SNR). Head diffraction is frequency dependent because high-frequency sounds with wavelengths smaller than the size of the head are more easily blocked. High-frequency attenuation can be as great as 20 dB and low-frequency attenuation is approximately 3 to 6 dB in the normal binaural auditory system. Binaural squelch refers to the central auditory nervous system's ability to compare the SNRs between the two ears and then use the noise from the ear with the poorer SNR to



AUDIOLOGICAL RECORD

NAME AG

TEST DATE 5/25/2011

DOB 3/12/1944 SEX Female

AUDIOLOGIST AB

AUDIOMETER GSI 61

EARPHONE Conventional

Effective Masking Level in dB	RE in										AC/BC
	LE	60 / 70	60 / 75		60 / 75		60 / 70	60 / 75	60 / 75	60	60	AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT		NR		DNT			
LT		15		100%	60		

Word Recognition List NU-6 R DNT L 2A R L

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	110	100	110	ABS	--	--
IPSI (X)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	80	75	90	100		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.4	1.3

Static Admittance (mL): Rt 0.4 Lt 0.4

Peak Pressure (daPa): Rt 10 Lt 15

Type: Rt A Lt A

COMMENTS: Right bone conduction threshold at 250 Hz was a vibrotactile response.

Fig. 44.1 AG's recent audiological examination.

partially remove the noise from the ear with the better SNR. Finally, binaural redundancy refers to the central auditory nervous system's ability to compare the speech inputs between the two ears and combine the inputs for a better representation of the message.

Interaural differences in time and intensity allow for the localization of sound. If sound reaches the right ear before the left ear, the person will likely localize the sound to the right side. Also, due to head diffraction, the level of sound is reduced when the sound bends around the head to reach the opposite ear. As a result, if the sound is louder in the right ear than the left ear, the sound will likely be localized to the right side.

UHL can severely affect a person's ability to recognize speech in noisy environments, especially if the noise is on the side of the better ear and the speech is on the side of the poorer ear. Also, localization can be very difficult because interaural differences in time and intensity are not available due to the total lack of hearing in one ear. If hearing is better in the right ear, then it is likely that sound will always be localized to the right side. Finally, high-frequency hearing loss in the better ear can further impact the person's ability to recognize speech in noisy environments. High-frequency hearing loss affects a person's ability to hear the high-frequency consonant sounds of speech and therefore decreases the overall clarity of speech.

2. What treatment options are available for UHL and which option would be most appropriate for this patient with a mild to moderate high-frequency hearing loss in the better ear?

Several amplification options are available in the treatment of UHL, including SoundBite (Sonitus Medical, San Mateo, CA), TransEar (Ear Technology Corp., Johnson City, TN), auditory osseointegrated implant system (AOIS), contralateral routing of signal (CROS), or bilateral contralateral routing of signal (BICROS). All of these options route sound from the poorer ear to the better ear. In other words, hearing is not restored in the poorer ear, and sound is not heard by the poorer ear. Instead, the sound is transferred either via bone conduction (BC) or a wireless electric signal to the better ear. Cochlear implantation (CI) in cases of UHL is also becoming an increasingly popular option, especially in Europe. CI would provide stimulation to the poorer ear and not route sound from the poorer ear to the better ear. CI, however, is not currently a standard option in the treatment of UHL and is not currently approved by the U.S. Food and Drug Administration (FDA) for use with UHL.

The SoundBite Hearing System routes sound from the poorer ear to the better ear via BC. With SoundBite, the patient wears a small behind-the-ear (BTE) transmitter on the poorer ear and a small removable custom-made retainer in the mouth. The BTE transmitter is a conventional open-fit hearing aid except the microphone is located in the ear canal. The custom-made retainer in the mouth contains an FM receiver, rechargeable battery, and oscillator. The retainer is strategically placed over the back molars on the side of the better-hearing ear. Sound is picked up by the microphone located in the poorer ear and wirelessly transmitted to the custom-made retainer in the mouth. The oscillator in the retainer then produces sound vibrations based on the signal from the microphone transmitter, which are conducted via the teeth, through the bone, to the cochlea of the better-hearing ear.

The TransEar also routes sound from the poorer ear to the better ear via BC. The TransEar is a conventional BTE hearing aid that is worn over the poorer ear. A custom earmold is fabricated that has a small BC oscillator encased in the shell. The earmold fits very deep in the ear canal so the oscillator makes contact with the bony portion of the ear canal. Sound is picked up by the microphone of the BTE hearing aid and is transmitted to the BC oscillator in the earmold via a thin wire. This signal to the oscillator causes the oscillator in the earmold to vibrate. The vibrations from the oscillator are transferred via the bones of the skull to the cochlea of the better ear. The TransEar has two programs, one for quiet listening environments and one with adaptive noise reduction for noisy listening environments. A directional microphone option is also available in the TransEar.

The AOIS is an osseointegrated device that routes sound from the poorer ear to the better ear via BC. A titanium screw with an abutment is implanted in the mastoid bone behind the poorer ear and osseointegrates with the bone. Typically, 3 months is required for the screw to osseointegrate with the bone before a small sound processor is snapped onto the abutment. The sound processor's microphone picks up sound, and the processor converts the sound to mechanical vibrations. The vibrations are then transferred to the cochlea of the better ear via BC. The auditory osseointegrated implant system was first introducted by Cochlear Americas and has been in use for over 30 years with patients with conductive and mixed hearing losses. It was introduced for use with patients with UHL in 2002. Recent advances, such as automatic multichannel adaptive directional microphones and noise reduction, allow for improved hearing in noise.

With CROS or BICROS amplification, the patient wears an amplitude-modulated (AM) or frequency-modulated (FM) microphone on the poorer ear, which converts the incoming sound to an electrical signal. The electrical signal is transmitted to an FM receiver on the better ear. When first introduced, the transmitter and receiver were connected via a wire. Currently, there are at least two manufacturers, Unitron (Plymouth, MN) and Phonak (Warrenville, IL), with wireless CROS and BICROS amplification. Typically, a hearing aid is used as the receiver. With the CROS, the hearing in the better ear is normal; therefore, the sound is simply routed from the poorer ear to the better without any modification of the sound. The hearing aid receiver is then anchored in the ear using a thin-tube and open dome or a standard earmold with a large open vent. With the BICROS, some magnitude of hearing loss is present in the better ear. Therefore, the hearing aid is appropriately fit to provide adequate amplification to the better ear, and then sound is also routed from the poorer ear to the hearing aid in the better ear. In BICROS amplification, microphones are active on both ears, and the signal from both microphones is modified based on the programming in the hearing aid receiver on the better ear. The wireless Phonak CROS and BICROS amplification available now uses directional microphone technology on the receiver side to improve the recognition of speech in noisy environments, whereas the Unitron CROS and BICROS amplification utilizes noise reduction on the transmitter and receiver sides.

Although AG is a candidate for all the devices just discussed, only the BICROS option allows for the optimization of the hearing loss present in the better ear. BICROS amplification will provide high-frequency amplification in the better left ear while also transmitting sound from the poorer right ear. The BICROS would be recommended for AG due to the high-frequency hearing loss in the better left ear.

3. How would a clinician counsel the patient on the realistic expectations of options for amplification for UHL?
It is important for the patient to understand that amplification options for UHL will not restore the hearing in the poorer ear. All sound is still routed to the better ear and therefore, only heard by the better ear. The options already discussed will allow patients to hear talkers on their poorer side, but the benefits of binaural hearing will not be restored. Localization of sound and recognizing speech in the presence of noise are both difficult for patients with UHL who do not use amplification, and they will continue to be difficult with the amplification options available for UHL. Typically, a patient with UHL will perform very well in a situation where noise is on the side of the poorer ear and the signal is on the side of the better ear. With the amplification options for UHL, however, routing sound from the poorer ear to the better ear, the noise from the side with the poorer ear will now be mixed with the signal to the better ear causing possible distraction from the signal. On the contrary, when noise is present on the side of the better ear and the signal is on the side of the poorer ear, the amplification options for UHL will be very helpful. It is important for patients with UHL to understand the benefits and limitations of the amplification options.
4. What type of verification is needed to fit a BICROS device?
The first step is to ensure that the hearing aid receiver is working correctly and programmed appropriately using real-ear measurements. All other measurements are based on the assumption that the signal processing of the hearing aid on the receiver side (i.e., better ear) provides appropriate gain and output.

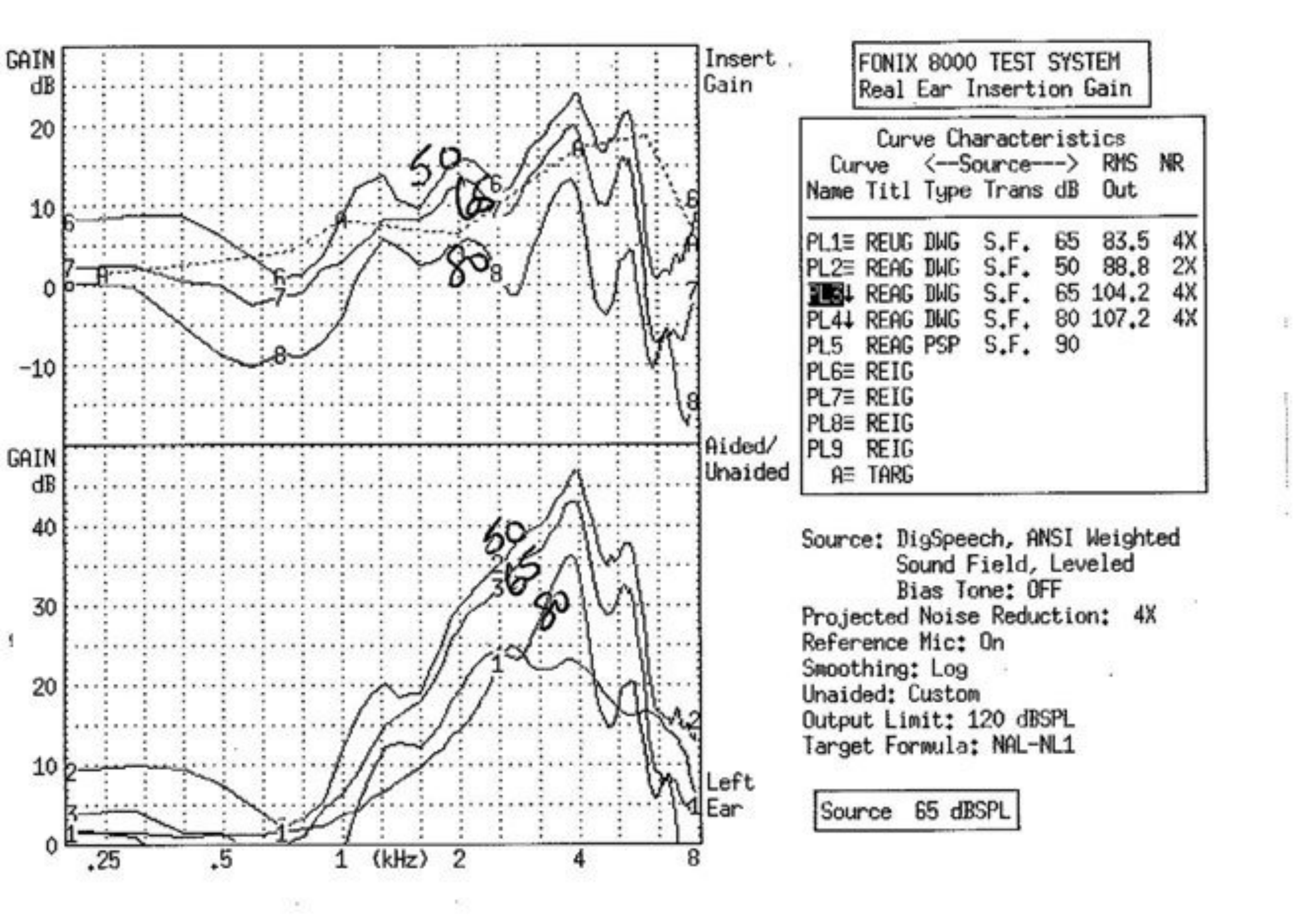
After it has been determined that the hearing aid receiver is working appropriately, transparency with the BICROS transmitter can be verified in the hearing aid analyzer. The output of the receiver hearing aid alone is measured in the hearing aid analyzer using a digital speech signal at an input level of 65 dB sound pressure level (SPL). The BICROS transmitter should be turned off during this first measurement. The BICROS transmitter is then turned on and placed in the test box of the hearing aid analyzer. The hearing aid, attached to the 2 cc coupler and test microphone, is placed on top of the test box on a foam pad. The output of the hearing aid when coupled to the BICROS transmitter is then measured once again using a digital speech signal at an input level of 65 dB SPL. The frequency response and output of the hearing aid coupled to the BICROS transmitter should be similar to that of the hearing aid alone. If the output of the hearing aid coupled to the BICROS transmitter is significantly different from that of the hearing aid alone then the volume of the BICROS transmitter should be adjusted.

44.5 Diagnosis and Recommended Treatment

All the different amplification options for UHL were discussed with AG. She was counseled that, because her hearing loss is mild to moderate in the high frequencies, she is a candidate for all the devices, including TransEar, AOIS, SoundBite, and BICROS. An AOIS was simulated on the right side by coupling the AOIS to a headband. AG liked the sound quality of the device but was not interested in having surgery. AG also expressed no interest in the TransEar due to fit issues. She was concerned that the deep fit of the TransEar would be uncomfortable. Finally, after much discussion, the SoundBite was ruled out as an option because AG was concerned about the dental work she has had on her back molars. As a result, BICROS devices were discussed in great detail. AG was also counseled that BICROS amplification was the best option for her due to her mild to moderate high-frequency hearing loss in the better left ear. It was discussed that ideal candidates for TransEar, AOIS, and SoundBite have completely normal hearing in the better ear. With the TransEar, AOIS, and SoundBite, there would be concern that she would no longer receive benefit if the hearing in her left ear declined any further.

AG was counseled that the left BICROS allows for amplification in the high frequencies to optimize the hearing in the left ear and then also route sound from the BICROS transmitter on the right side. The Unitron Tandem 16 and the Phonak BICROS hearing aids were discussed. She was also counseled that the devices use either noise reduction or directional microphones to improve the recognition of speech in background noise. AG chose the Phonak BICROS, using an Audeo S Smart III hearing aid (Phonak) for the left ear and the BTE CROS transmitter with a 312 battery on the right ear.

The left Phonak Audeo S Smart III aid was fit using an open dome due to her normal hearing from 500 to 2000 Hz in the left ear. Real ear insertion gain (REIG) targets corrected for six channels of signal processing was established using the National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) fitting rationale, and real-ear measurements for input levels of 50, 65, and 80 dB SPL were performed. Measurements revealed appropriate gain for the three input levels and smooth frequency response (Fig. 44.2). The dashed curve ("A") represents the NAL-NL1 prescriptive REIG target for 65 dB SPL. Then transparency of the CROS transmitter was measured in the hearing aid analyzer test box (Fig. 44.3). First, an output curve was performed with the left hearing aid in the test box of the hearing aid analyzer and the CROS transmitter turned off (Curve 1 in Fig. 44.3). The CROS transmitter was then turned on and placed in the test box of the hearing aid analyzer. The hearing aid, coupled to the 2 cc coupler and test microphone, was placed on top of the test box on a foam pad and another output curve was measured (Curve 2 in Fig. 44.3). The output curves of the hearing aid measured alone (Curve 1) and the hearing aid with the CROS transmitter on (Curve 2) show similar frequency responses, and the average output of the hearing aid with the CROS transmitter (95.2 dB SPL) is 5.8 dB greater than the average output of the hearing aid alone (89.6 dB SPL).



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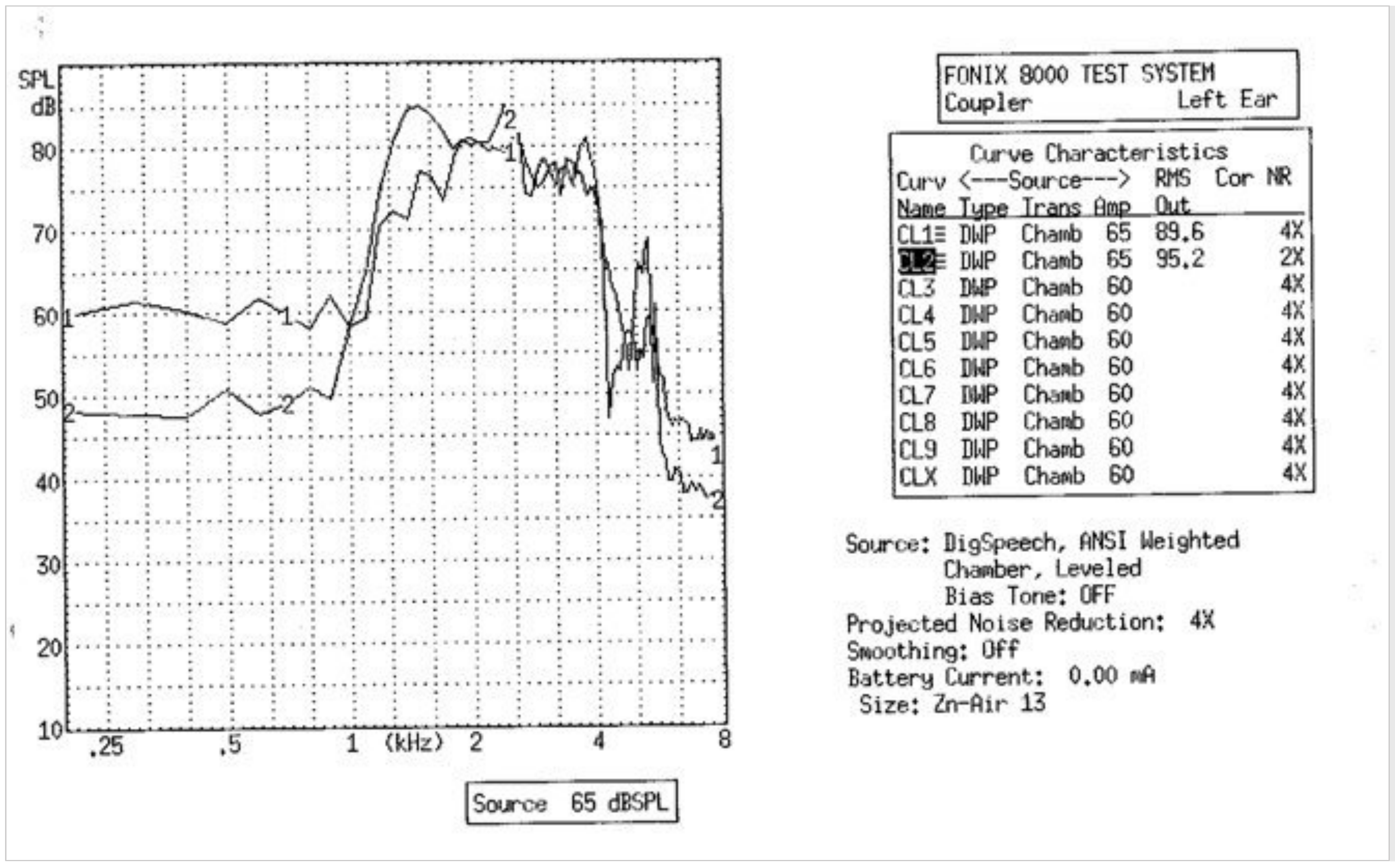
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

         Curve Characteristics
  Curve    <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1≡ REUG DWG  S.F.  65  83.5  4X
PL2≡ REAG DWG  S.F.  50  88.8  2X
PL3↓ REAG DWG  S.F.  65 104.2  4X
PL4↓ REAG DWG  S.F.  80 107.2  4X
PL5  REAG PSP  S.F.  90
PL6≡ REIG
PL7≡ REIG
PL8≡ REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL
```

Fig. 44.2 Real-ear measurements with AG's left Phonak Audeo S Smart III aid using the Frye 8000 real ear analyzer (Frye Electronics, Inc., Tigard, OR). Curve 1 is real ear unaided gain (REUG). Curve 2 is real ear aided gain (REAG) for a 50 dB SPL input. Curve 3 is REAG for a 65 dB SPL input and curve 4 is REAG for an 80 dB SPL input. Curve 6 is real ear insertion gain (REIG) for a 50 dB SPL input. Curve 7 is REIG for a 65 dB SPL input, and curve 8 is REIG for an 80 dB SPL input. The A curve is the NAL-NL1 target for a 65 dB SPL input.



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FONIX 8000 TEST SYSTEM
Coupler              Left Ear

        Curve Characteristics
Curv <---Source---> RMS  Cor NR
Name Type Trans Amp  Out
CL1≡ DWP  Chamb  65  89.6     4X
CL2≡ DWP  Chamb  65  95.2     2X
CL3  DWP  Chamb  60           4X
CL4  DWP  Chamb  60           4X
CL5  DWP  Chamb  60           4X
CL6  DWP  Chamb  60           4X
CL7  DWP  Chamb  60           4X
CL8  DWP  Chamb  60           4X
CL9  DWP  Chamb  60           4X
CLX  DWP  Chamb  60           4X

Source: DigSpeech, ANSI Weighted
        Chamber, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Smoothing: Off
Battery Current:  0.00 mA
 Size: Zn-Air 13
```

Fig. 44.3 Verification of CROS transmitter transparency on the Frye 8000 hearing aid analyzer Frye Electronics, Inc., Tigard, OR). Curve 1 is the output curve measured with the left hearing aid in the hearing aid analyzer test box and the CROS transmitter off. Curve 2 is the output curve measured with the CROS transmitter turned on and placed in the test box of the hearing aid analyzer and the hearing aid, coupled to the 2 cc coupler and test microphone, placed on top of the test box on a foam pad. The output curves of the hearing aid measured alone (curve 1) and the hearing aid with the CROS transmitter on (curve 2) show similar frequency responses and the average output of the hearing aid with the CROS transmitter (95.2 dB SPL) is 5.8 dB greater than the average output of the hearing aid alone (89.6 dB SPL).

44.6 Outcome

AG found her Phonak left BICROS to be very beneficial. She reported she does not ask for repetition in conversation as often, and she likes that she can have a conversation with a speaker on her right side. AG also finds that noisy listening environments are still very noisy, but she states she is not as bothered by the noise when wearing her left BICROS and that she can follow the conversation with less fatigue. AG states that she still has difficulty localizing sound, but she understands that this will continue to be an issue. Overall, AG is very pleased with her Phonak left BICROS and is using it regularly.

44.7 Key Points

1. Hearing loss in one ear can significantly impact communication by eliminating the benefits of binaural hearing.
2. Amplification options for UHL do not restore hearing to the poorer ear and therefore do not restore the advantages of binaural hearing.
3. Several amplification options are available for UHL, and all the options should be discussed with the patient. The patient should be counseled extensively on the benefits and limitations of each option.
4. If hearing loss is present in the better ear, then the BICROS device is typically the best option because the BICROS allows for amplification in the better ear. With SoundBite, AOIS, and TransEar, sound is simply routed to the better ear via BC, but the better ear still has hearing loss.
5. Verification is required when fitting a BICROS, and this verification begins with ensuring that the hearing aid in the better ear is fit properly using real-ear measurements.

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45 High Frequency Gain-Dependent Word Recognition

Michael J. Cevette, Jamie M. Bogle, Sarah O. Holbert, Linsey Scheibler, and Jan Stepanek

Four patients were evaluated using conventional and high frequency enhanced speech.

45.1 Clinical History and Description

The Articulation Index (AI), revised to the Speech Intelligibility Index (SII), is a tool used to predict the portion of the speech signal that is audible to a patient with a specific hearing loss. The frequency region above 1000 Hz (Fig. 45.1) is reported to contain "5%of the power of speech," but contributes to "60%of the ability to recognize speech" and reflects the importance of high frequency audibility in word recognition (Fig. 45.1). As high frequency audibility increases, word recognition is generally expected to increase. This is particularly demonstrated in the presence of relatively intense background noise where increased high frequency information has been found to be beneficial in all cases regardless of the magnitude and configuration of hearing loss. For most listeners with normal hearing and for those with mild and moderate hearing loss, the AI or SII provides a good predictor of the impact of audibility on word recognition.

The extent of the benefit from restored high frequency audibility, however, has been controversial, particularly when considering severe high frequency sensorineural hearing loss. For example, limited improvement in word recognition with additional high frequency gain has been reported for patients with greater than a mild high frequency sensorineural hearing loss. This lack of improvement has been attributed to the presence of areas of the cochlea with nonfunctional inner hair cells and/or auditory neurons, termed "cochlear dead hair cell regions" (CDHCRs). Some investigators have shown that the presence of CDHCRs may limit accurate transmission of high frequency speech information with little to no improvement in word recognition even with the additional high frequency gain.

Because frequencies above 1000 Hz are relatively more important to word recognition than frequencies below 1000 Hz, audiologists may assess the impact of altering the presentation of word recognition lists by enhancing the high frequency gain. By amplifying the more highly weighted frequencies above 1000 Hz, the patient may display improved word recognition. Currently, there is no established clinical method for applying additional high frequency gain during conventional word recognition testing in order to determine the influence of audibility on word recognition performance. These four case reports demonstrate the effect of providing 14 dB of additional gain above 1000 Hz upon word recognition scores (WRSs) in quiet and discuss the consequent impact on recommendations regarding amplification and diagnostic testing using word recognition materials.

45.2 Audiological Testing

Four adults (Fig. 45.2 and Fig. 45.3; patients A–D) with high frequency (>1000 Hz) hearing thresholds greater than 55 dB HL and WRSs of 80% or poorer are described. Word recognition testing was conducted using recorded phonemically balanced consonant-vowel-consonant (CVC) monosyllabic word lists ("English Speech Audiometry," Brigham Young University, 1998). Each word list contains 10 words, and two lists were provided for each of the two (i.e., original and high frequency gain-adjusted) test conditions. The initial and final consonants appear in accordance with their frequency of use in these positions. The presentation level for each case was adjusted based on the patient's hearing threshold levels.

After audiometric threshold and baseline word recognition testing were completed, 14 dB of gain at all frequencies above

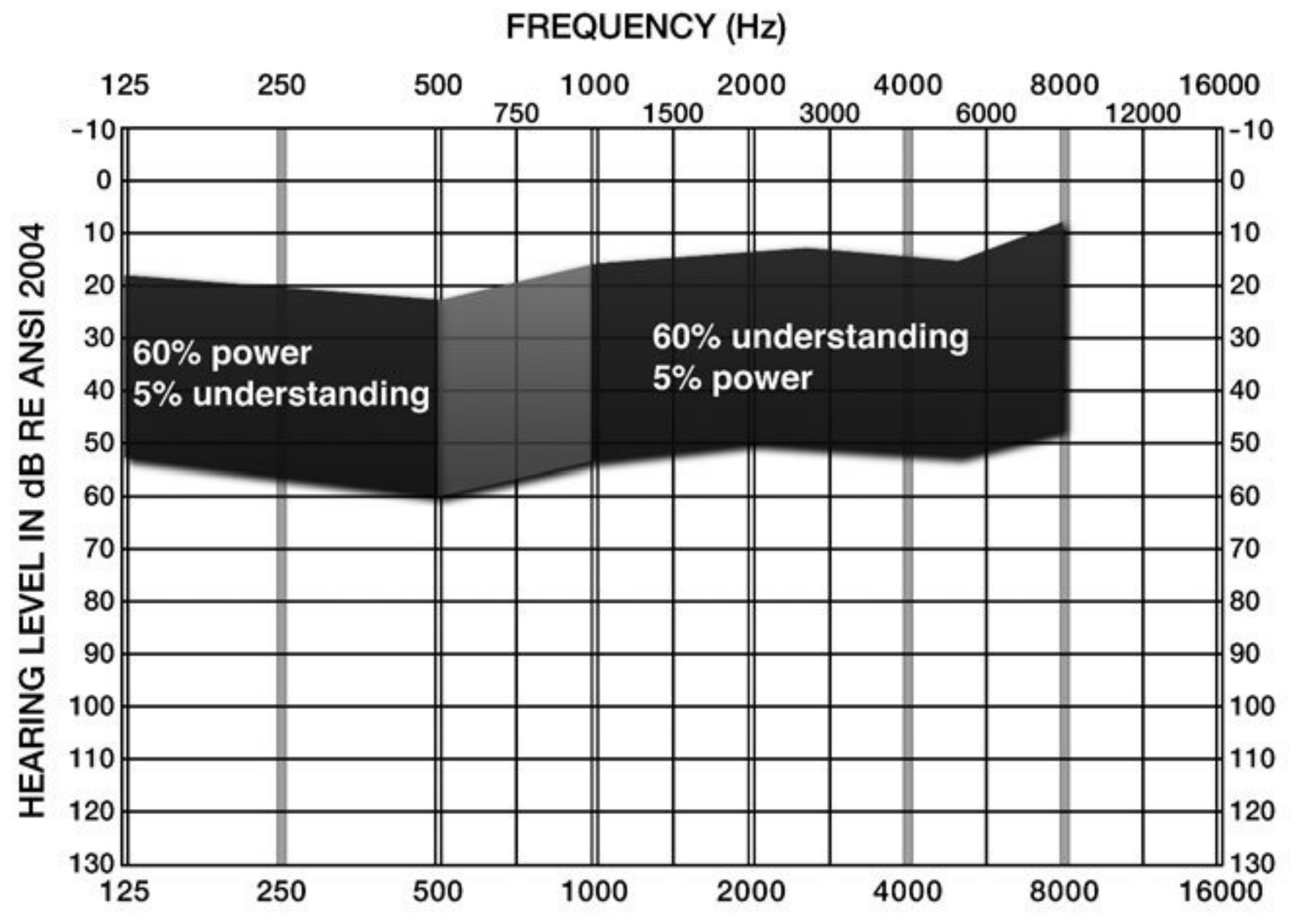

Fig. 45.1 The displayed speech spectrum illustrates the relative proportion of power and speech understanding below 500 Hz and above 1000 Hz.

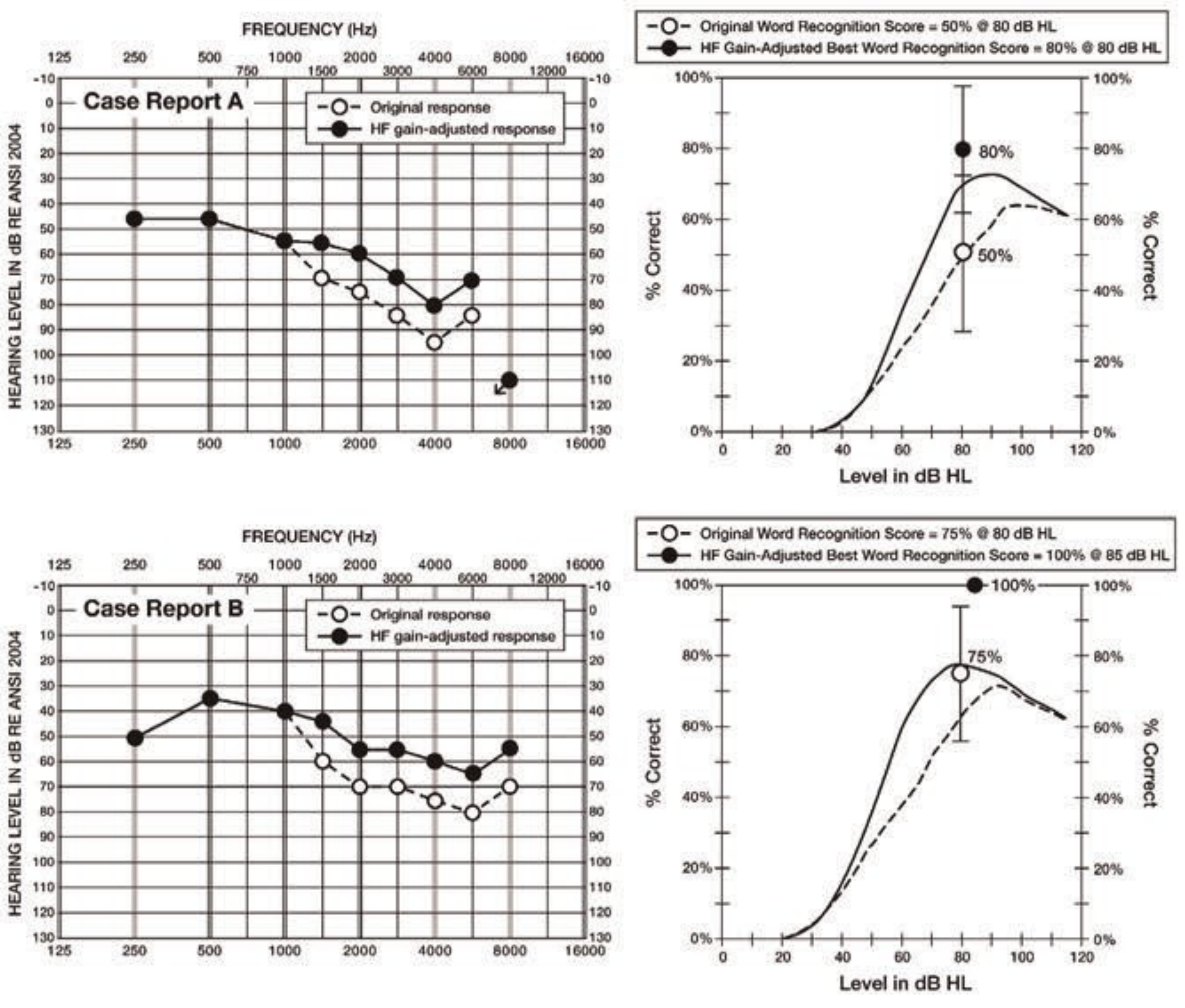

Fig. 45.2 Original pure-tone thresholds and word recognition scores (WRSs) are in open circles. Closed circles indicate thresholds and WRSs obtained using a 14 dB high frequency gain-adjusted filter. The PI-PB function to the right reports that the WRSs were compared with clinically obtained normative data for this degree of hearing loss. The dashed line describes the expected WRSs for the obtained hearing loss, whereas the solid line represents expected scores for thresholds obtained using additional high frequency gain. Both cases demonstrate mild to moderate sensorineural hearing loss. For Case A additional high frequency gain provided a 30% improvement in WRS, whereas for Case B the improvement was 25%

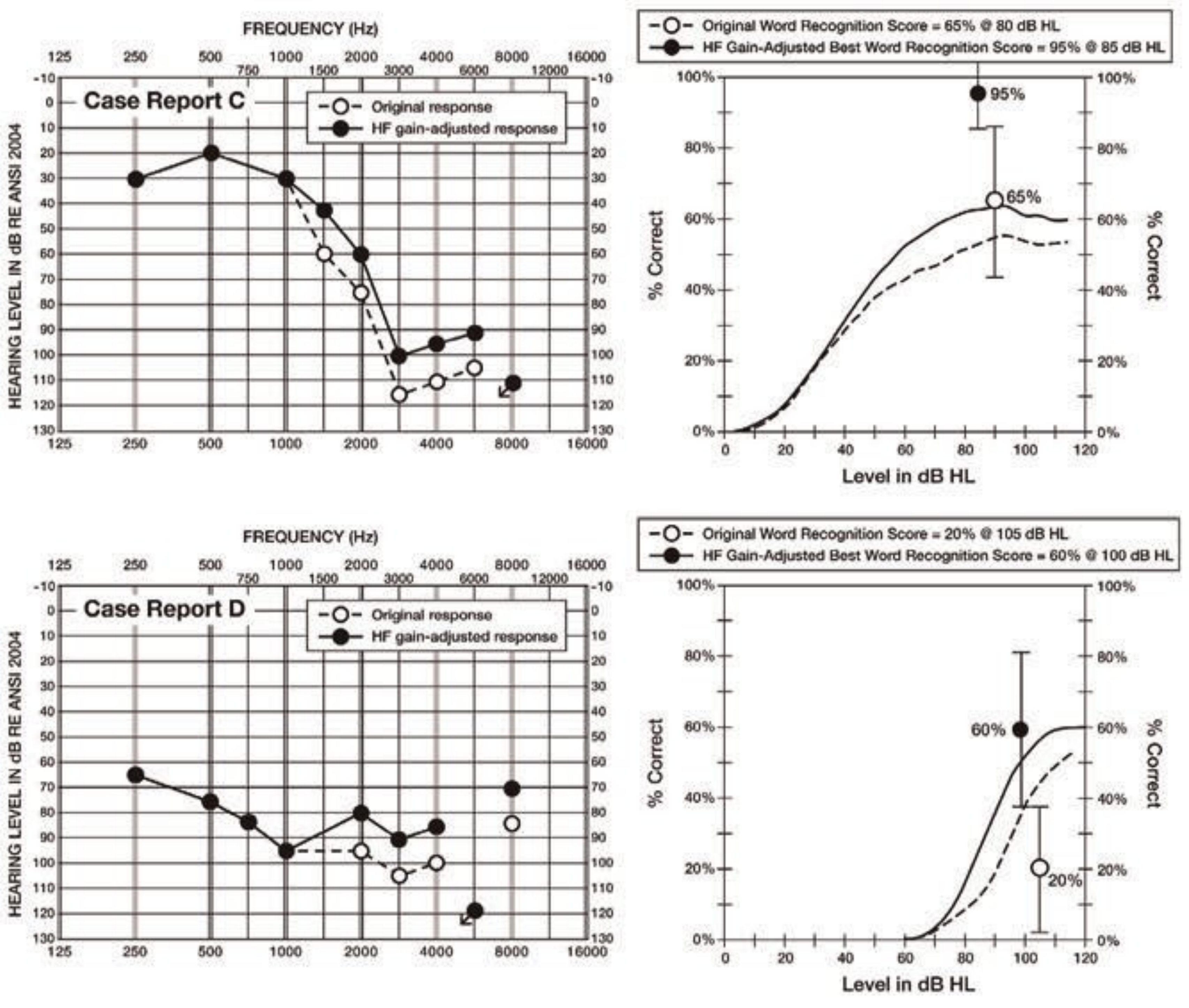

Fig. 45.3 Original pure-tone thresholds and word recognition scores (WRSs) are in open circles. Closed circles indicate thresholds and WRSs obtained using a 14 dB high frequency gain-adjusted filter. The PI-PB function to the right reports that the WRSs were compared with clinically obtained normative data for this degree of hearing loss. The dashed line describes the expected WRSs for the obtained hearing loss, whereas, the solid line represents expected scores for thresholds obtained using additional high frequency gain. For Case C additional high frequency gain provided a 30% improvement in WRS, whereas for Case D the improvement was 40%

1000 Hz was applied. This was the maximum available output for the sound processor (Ultra-Curve Pro digital sound processor, Model DEQ2496, Behringer, Chesterton, IN) used for these four patients. This processor was purchased off-the-shelf and was connected to a Grason-Stadler 61 diagnostic audiometer (Grason-Stadler, Eden Prairie, MN). No additional software or hardware was required. Next, additional lists of high frequency gain-adjusted filtered words were then presented. For Cases A, B, and C, presentation level was set to 80 to 85 dB HL, which provided comfortably audible sensation levels (27–48 dB sensation level [SL] relative to pure-tone average). The hearing thresholds for Case D were more significant (pure-tone average = 88 dB HL), requiring a higher presentation level (i.e., 100 dB HL) to provide audibility. Original and high frequency gain-adjusted audiograms and the resulting WRSs are reported in Fig. 45.2 and Fig. 45.3. Fig. 45.2 reports two cases (Cases A and B) with similar mild to moderate sensorineural hearing loss, but with different “original” and “high frequency-gain adjusted” WRSs. Fig. 45.3 reports the results for one patient with mild to profound sensorineural hearing loss (Case C) and another patient with severe sensorineural hearing loss (Case D).

The addition of 14 dB of gain at above 1000 Hz led to improvement in WRSs. For Case A (Fig. 45.2a), the WRS improved from 50 to 80% with the addition of high frequency gain. Similarly, for Case B (Fig. 45.2b), WRS improved from 75 to 100% For these two cases, good high frequency-gain adjusted WRSs were obtained. Fig. 45.3 a provides an "original" WRS of 65% with an improvement to 95% with the additional 14 dB of high frequency gain. Finally, Fig. 45.3b describes a case of a severe sensorineural hearing loss with a poor "original" WRS of 20% The addition of 14 dB of high frequency gain improved the WRS to 60% In these four cases, the addition of 14 dB of high frequency gain altered the prediction of the inherent cochlear distortion associated with the degree and configuration of the sensorineural hearing loss. This method provides additional clinical information that may lead to improved counseling with patients regarding hearing aid benefit. That is, will the addition of high frequency gain provide improved performance?

45.3 Questions to the Reader

1. Describe the performance intensity function for word recognition testing. How is it used as a diagnostic tool for site of lesion testing?
2. Describe the theory of CDHCRs. How might this impact speech recognition?
3. Is speech recognition testing a helpful measure in counseling patients regarding the potential benefits of amplification?

45.4 Discussion of Questions to the Reader

1. Describe the performance intensity function for word recognition testing. How is it used as a diagnostic tool for site of lesion testing?
   Performance intensity (PI) functions describe word recognition ability as a function of increased presentation level. Clinically, PI functions have been used to estimate the degree of auditory distortion as an alternate test for site of lesion. Phonemes typically become more easily identified as the presentation level increases and audibility is improved. WRSs range from 0% to maximum performance using phonetically balanced word lists (termed PB^{max}) for phonemically balanced words. Clinically, PI functions have been used to differentiate between cochlear and retrocochlear lesions. WRSs for individuals with normal hearing or cochlear hearing loss often plateau with increasing presentation levels; however, those with retrocochlear lesions have demonstrated decreased WRSs as the presentation level increased (rollover index (RI) = $PB_{max} - PB_{min}/PB_{max}$). PB_{min} is defined as the lowest score obtained at a presentation level greater than the intensity used to obtain PB_{max}. Rollover is considered clinically significant if the RI is > 0.45. PI functions are calculated by increasing the presentation level, but measuring this function may be limited due to loudness discomfort. As demonstrated in these four cases, additional improvement in WRS may be obtained by increasing only the presentation level at frequencies above 1000 Hz. Although it is unclear how this will impact site of lesion estimations, this method provides additional information regarding WRSs and possible outcomes when using amplification.
2. Describe the theory of CDHCRs. How might this impact speech recognition?
   CDHCRs are areas of the cochlea that lack functioning inner hair cells and/or neurons. These areas are physiologically incapable of transmitting information along the auditory pathway. Unfortunately, these regions are not always apparent with audiometric testing. According to the CDHCR, auditory input may be detected by the spread of excitation to a functional area. This leads to the occurrence of auditory perception at nonappropriate frequencies (i.e., at frequency regions not originally assigned to analyze the incoming signal). For example, if a patient has a CDHCR in the area of 1500 Hz, the psychometric tuning curve will provide stimulation at additional frequencies along the basilar membrane including 1000 Hz. The audiologist may misinterpret this patient's test results because of this altered detection. For patients with CDHCRs, adding additional high frequency information through amplification may or may not be beneficial, as suggested by studies demonstrating variable improvement in WRS following hearing aid fitting in patients with moderate to severe sensorineural hearing loss. On the other hand, other studies have indicated similar improvements in WRS in quiet between patients with and without the described CDHCR. A difference in performance was found when testing in conditions with noise, suggesting that patients with CDHCR may improve performance in quiet conditions but may experience more difficulty in the presence of background noise.
3. Is speech recognition testing a helpful measure in counseling patients regarding the potential benefits of amplification?
   Speech recognition testing can provide helpful information regarding how a patient will perform with amplification, especially for patients with "poor" or "excellent" WRSs. Patients within the "excellent" or "ceiling range" of speech recognition performance typically perform well with amplification, and patients within the "poor" or "floor range" of speech recognition performance typically may not achieve significant benefit. For patients within the "fair" or "good" performance range of speech recognition performance, the outcome is often less predictable, and the assumption that word recognition performance is an index of cochlear distortion may not be completely valid. Certainly, these four cases demonstrate that some patients with sloping high frequency sensorineural hearing loss have improved performance with increased high frequency audibility. Ultimately, this may lead to implications regarding counseling and use of amplification.

45.5 Diagnosis and Recommended Treatment

The improvement in word recognition performance with the additional 14 dB of gain above 1000 Hz when compared with conventional word recognition testing provides useful clinical information. Although this limited number of patients may not translate to a larger population, the findings do suggest that there are useful implications for using this approach for diagnostic use of word recognition in a clinical setting. This is particularly true when used as an indication of speech distortion or in counseling as a marker for the potential benefit from amplification.

45.6 Outcome

For these four cases, there was significant improvements in WRSs when baseline WRSs were measured at 80% or poorer. These findings may have relevance for the method of testing WRS and for counseling potential hearing aid users.

45.7 Key Points

1. Additional test measures may be useful for predicting potential benefit with amplification, particularly for patients with moderate, severe, and profound sensorineural hearing losses.
2. Improvement in WRSs that is dependent upon high-frequency audibility is reflected in the AI or SII function
3. When applying additional high frequency gain, the performance-intensity rollover index may be affected.

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46 The Case of the Missing C#

Marshall Chasin

HB is a 68-year-old man who reports he no longer enjoys playing his piano and feels the notes around C# on his piano sound slightly "flat."

46.1 Clinical History and Description

HB was referred by his family physician to an audiology clinic specializing in music and related issues. HB is 68 years old and is an amateur piano player. HB has worked in a range of noisy environments and is an avid gun enthusiast. There are no other otologic or audiological issues reported by the patient. HB has always enjoyed music and because of his recent retirement is playing the piano more frequently. HB reported some distortion whenever he played C# on his piano. When questioned further, HB reported that the note C# on his piano tended to sound slightly "flat." HB had his piano recently tuned and still notices this poor sound quality. HB also reported that the higher-pitched notes on the right side of the piano keyboard sound "thin."

46.2 Audiological Testing

A comprehensive audiological examination (Fig. 46.1) was completed with a suspicion of presbycusis and noise exposure. Tympanometry indicated normal middle ear function with ear canal volume, static compliance, and middle ear pressure all within normal limits bilaterally. Contralateral and ipsilateral acoustic reflex thresholds (ARTs) were elevated bilaterally at 500 and 1000 Hz and absent at 2000 and 4000 Hz. This latter finding would be expected given the magnitude of hearing loss present at 2000 and 4000 Hz. Using ER-3A insert earphones (Etymotic Research, Inc., Elk Grove Village, IL), audiometric

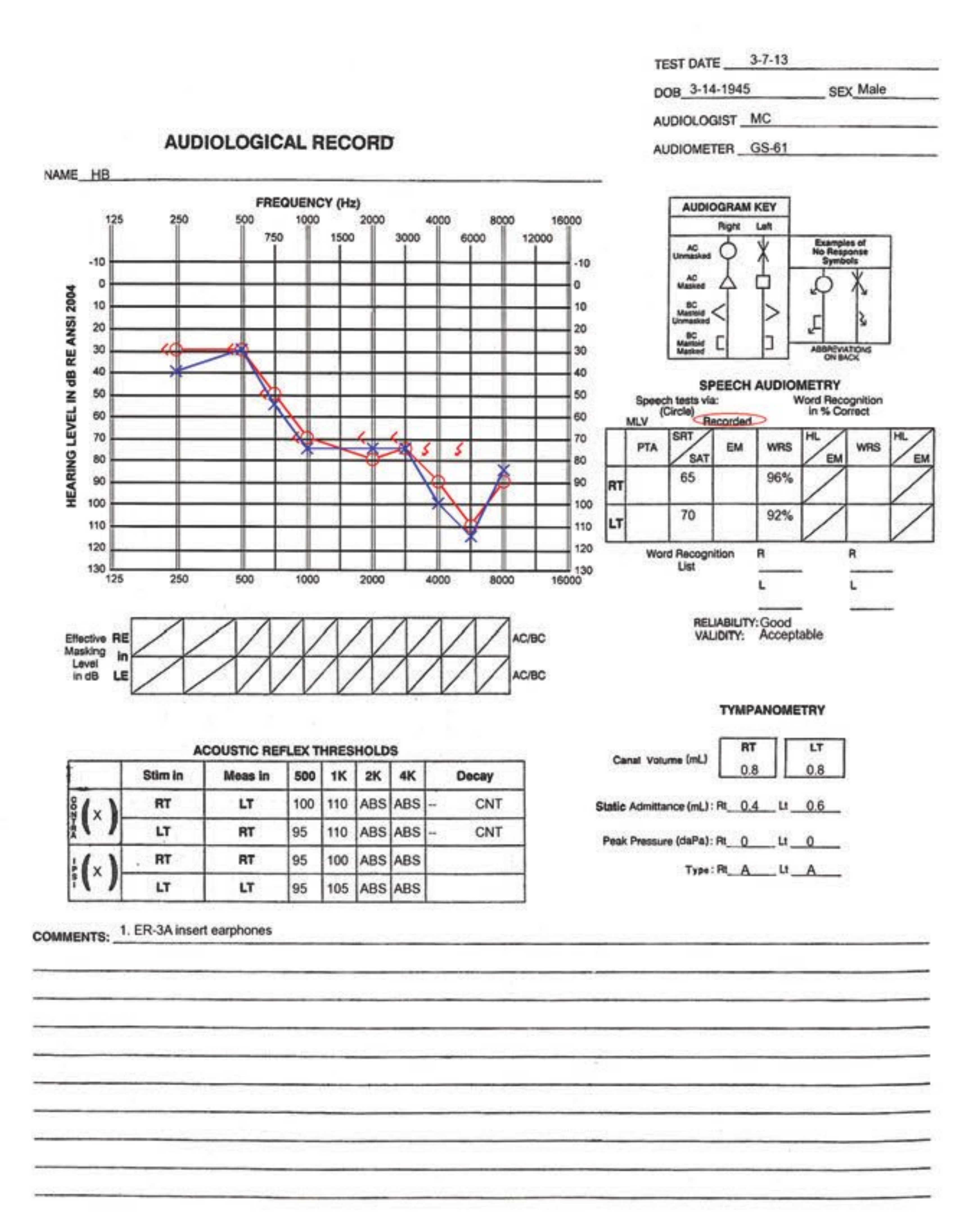

Fig. 46.1 Audiological examination revealing the bilateral symmetrical sensorineural hearing loss for HB.

AUDIOLOGICAL RECORD

TEST DATE 3-7-13
DOB 3-14-1945 SEX Male
AUDIOLOGIST MC
AUDIOMETER GS-61
NAME HB

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT		65		96%			
LT		70		92%			

Word Recognition List R ___ L ___ R ___ L ___

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay
CONTRA (X)	RT	LT	100	110	ABS	ABS	-- CNT
	LT	RT	95	110	ABS	ABS	-- CNT
IPSI (X)	RT	RT	95	100	ABS	ABS	
	LT	LT	95	105	ABS	ABS	

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.8	0.8

Static Admittance (mL): Rt 0.4 Lt 0.6
Peak Pressure (daPa): Rt 0 Lt 0
Type: Rt A Lt A

COMMENTS: 1. ER-3A insert earphones

thresholds revealed a bilateral symmetrical mild to profound bilateral symmetrical high-frequency sensorineural hearing loss that is sharply falling in configuration beyond 500 to 1000 Hz and is flat to 3000 Hz, then sharply falling once again to 6000 Hz with some recovery to 8000 Hz. Speech recognition thresholds (SRTs) revealed a moderate to severe loss bilaterally in the ability to receive speech. Word recognition scores (WRSs) revealed normal ability (96% in the right ear and 92% in the left ear) to recognize speech bilaterally. Otoacoustic emission (OAE) testing revealed no measureable responses bilaterally.

It was explained to HB that he had bilateral symmetrical high-frequency sensorineural hearing loss, which is consistent with his age (presbycusis) and his history of exposure to noise. It is expected that HB would likely have only slight difficulty communicating in a quiet, one-to-one environment, but he may experience significant difficulty recognizing speech in more adverse listening environments. In addition, this loss of high-frequency hearing (and the subsequent loss of harmonic audibility) may help explain HB's report that the higher-pitched notes on the piano sounded thin. At this point, bilateral hearing aids were recommended for social situations as well as when HB is playing or listening to music.

46.3 Questions to the Reader

1. Which phonemes would HB be expected to have greater difficulty hearing and what contribution would these missing phonemes provide to the Speech Intelligibility Index (SII) for an English-speaking listener?
2. Why might HB hear some notes as flat and not sharp?
3. What are some strategies that could be implemented in the clinic that could improve HB's enjoyment of music?

46.4 Discussion of Questions to the Reader

1. Which phonemes would HB be expected to have greater difficulty hearing and what contribution would these missing phonemes provide to the Speech Intelligibility Index (SII) for an English-speaking listener?
   The phonemes that HB would be missing would be the mid- and higher-frequency obstruent sounds such as the sibilants, affricates, and fricatives (s, z, ch, f, v). In addition, HB would be missing aspiration cues that are associated with all voiceless stops in English. Because the SII for English has greater weighting for the mid and high frequencies, many of these important sounds that contribute to the clarity of speech would not be audible. For the audiometric configuration shown in Fig. 46.1, the unaided SII is 14%, which implies a significant communication difficulty, especially in noisy environments. The SII was calculated by entering HB's audiogram into a real ear measurement system that automatically performs the calculation. This resulting SII suggests that only 14% of the available speech cues for correct speech recognition are available to HB because of the magnitude and configuration of his hearing loss.
2. Why might HB hear some notes as flat and not sharp?
   Hallowell Davis and his colleagues delineated the essential elements required for normal pitch perception in a series of experiments. When pitch perception is not normal, Davis et al referred to this phenomenon as "dyplacusis." In these experiments, American servicemen were asked to subject themselves to a monaural exposure of an intense level of noise for a period of time that resulted in temporary threshold shift (TTS). The opposite ear was protected from the exposure to noise. The servicemen were then asked to match loudness and pitch for pure-tones presented to the better ear to the same signal presented to the poorer ear. In the region of sensorineural hearing loss, as the probe tone frequency was gradually increased in the good ear, the participants judged the sound to be getting louder, but not higher in pitch. That is, the participants gradually heard the probe sound as louder, but "flatter" than what the probe frequency was. This phenomenon was judged to be of cochlear origin and is related to significant neural damage. It is most likely similar to the finding of a "dead hair cell region" for the cochlear inner hair cells as delineated by Moore et al. There is no known cochlear phenomenon that would cause a patient to hear sounds as being sharp.
3. What are some strategies that could be implemented in the clinic that could improve HB's enjoyment of music?
   With current technology there are two approaches that may offer some help for HB to again enjoy the music while he plays his piano. One is related to any number of frequency-shifting or frequency-transposing technologies that are currently commercially available where the offending input signal is shifted or transposed to a healthier portion of the cochlea. Another approach (used in this case report) is the simple removal (notch filtering) of the offending frequency region (or regions). Although neither of these approaches is optimal, over time, both approaches may significantly improve HB's enjoyment of music.

46.5 Optional Testing

Because there were no pitch perception tests that were routinely performed in the audiology clinic of this case report, which is a musicians' clinic, HB was escorted to the piano in the clinic. At this point, it should be mentioned that there are a range of inexpensive, downloadable software-based applications that will allow a clinician to perform some pitch-matching tests. While sitting at the piano, HB was asked to play the notes on the right-hand side of the keyboard starting with middle C [262 Hz] and to inform the clinician which notes were causing his report of difficulty with pitch perception. The reported notes were the three octave C# notes at 277, 554, and 1109 Hz. The higher-frequency C# note at 2218 Hz and the top note on the piano keyboard, high C at 4186 Hz, did not seem to cause a problem with pitch perception, although these notes were not as loud as HB thought they should be.

Although not performed, additional testing could also have included the threshold equalizing noise (TEN) test with calibration in dB HL to evaluate the existence and extent of any cochlear dead hair cell regions. The commercially available test has been used extensively to inform the clinician of potentially problematic frequency regions for a patient.

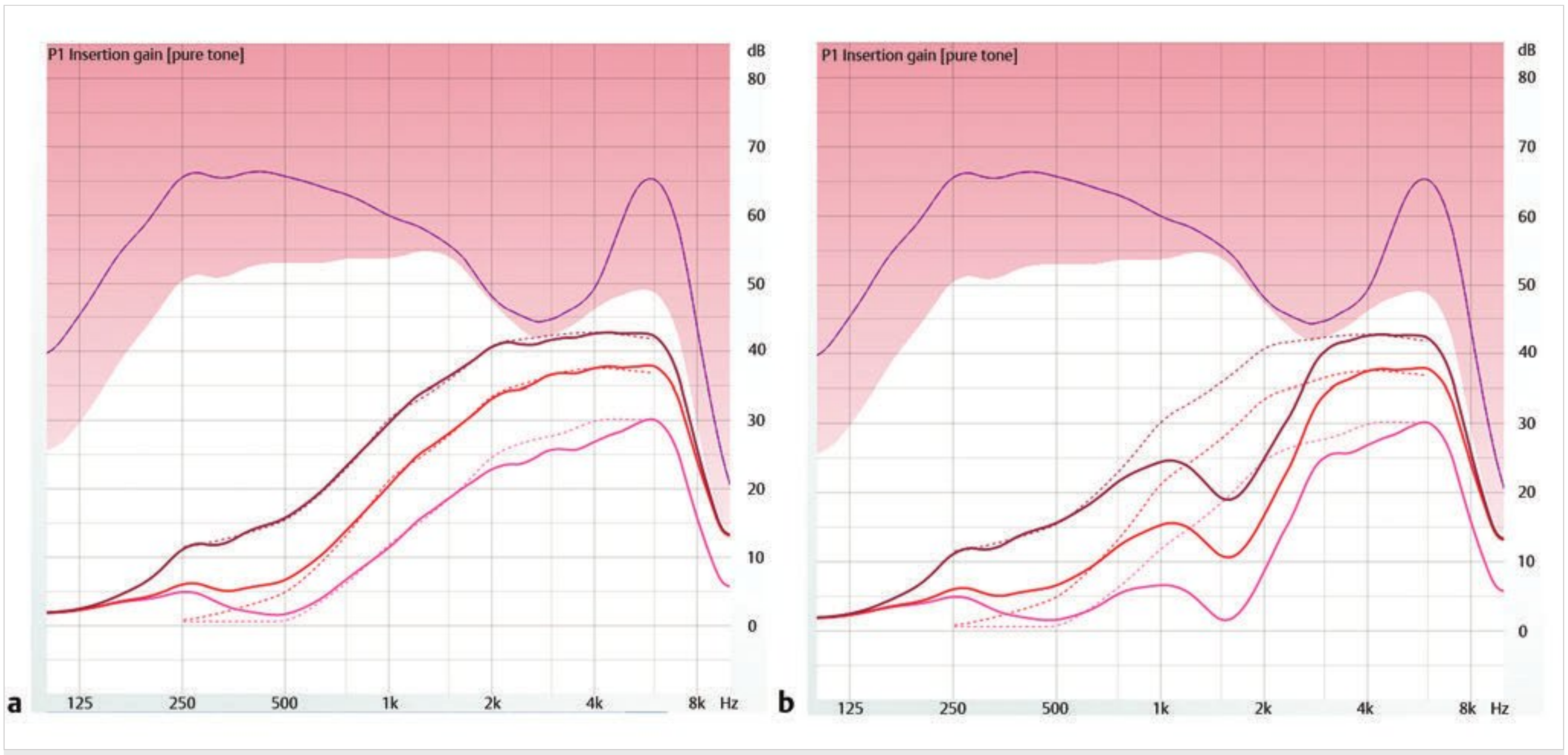

Fig. 46.2 (a) NOAH screen shot for the unfiltered insertion gain for soft (upper curve), average (middle curve), and loud (lower curve) input levels for the right ear only, using NAL-NL2 (dashed lines for the soft, average, and loud input levels). The top line reports the maximum amount of gain before feedback is expected to occur. (b) NOAH screen shot for the insertion gain with a 15 dB notch between 1000 and 2000 Hz for soft (upper curve), average (middle curve), and loud (lower curve) input levels for the right ear only, using NAL-NL2 (dashed lines for the soft, average, and loud input levels). The top line shows the maximum amount of gain before feedback is expected to occur.

46.6 Diagnosis and Recommended Treatment

HB had dyplacusis, or perhaps dead hair cell region(s) in his cochleae that is most likely related to significant inner hair cell (IHC) damage. Bilateral hearing aids with wide dynamic range compression (WDRC) were recommended and fit. These digital hearing aids had sufficient flexibility in their software to selectively reduce the gain in specific frequency regions. In Fig. 46.2 a, the three solid lines in the nonshaded area represent the initial measured real ear insertion gain (REIG) for soft (upper curve), average (middle curve), and loud (lower curve) input levels for the right ear only using the NAL-NL2 prescriptive target (i.e., the dashed lines for the soft, average, and loud input levels). The top line in the shaded area reports the maximum allowable gain before feedback might occur. As can be seen in Fig. 46.2a, the initial measured REIG to the NAL-NL2 targets for the three input levels was quite acceptable.

After this initial fit, the gain for the first C# in the 250 to 300 Hz region (C# [274 Hz]) was reduced by 15 dB, but HB did not report any improvement with that C# note as he played the piano. Suspecting that the offending region may be due to a higher-frequency harmonic of C# [274 Hz], reducing the REIG at around the next octave higher C# [554 Hz] was addressed. Again, when HB played the piano, he reported no significant subjective improvement when this frequency region was also reduced by 15 dB. Finally, the C# (1109 Hz), which is an additional octave higher, was addressed, and notch filtering of 15 dB in the region between 1000 Hz and 1500 Hz (Fig. 46.2 b) did substantially improve the sound quality of the piano (Fig. 46.2). As can be seen in Fig. 46.2 b, there is a 15 dB reduction in gain for the three input levels in the frequency region between 1000 and 2000 Hz. Using this relatively simple subjective approach, it appears that the frequency region of significant cochlear hair cell damage for HB was centered at approximately C# (1109 Hz), and any reports of poor sound quality of the lower-frequency C# note may have been due to the higher-frequency harmonics in the 1000 to 2000 Hz region.

46.7 Outcome

HB was able to successfully use his bilateral amplification and reported his piano sounded significantly better, albeit not perfect. HB was able to use the hearing aids in many social situations with significant benefit.

46.8 Key Points

1. A report of a musical note sounding flat may be an indicator of significant cochlear damage, which may be related to a cochlear dead hair cell region.
2. Avoiding the offending frequency region (either by frequency transposition or by gain reduction) may improve sound quality.
3. In music, a report about a specific frequency region may be related to a higher-frequency harmonic rather than the fundamental (or note name).

Suggested Reading

Chasin M. The case of the missing C#. Hear Rev 2012; 19: 16–19

Davis H, Morgan CT, Hawkins JE, Galambos R, Smith FW. Temporary deafness following exposure to loud tones and noise. Acta Otolaryngol Suppl 1950; 88 Suppl.88: 1–56

Moore BC, Glasberg BR, Stone MA. New version of the TEN test with calibrations in dB HL. Ear Hear 2004; 25: 478–487

47 Do You Hear What I Hear?

Jackie L Clark

MT is an experienced hearing aid wearer reportedly receiving radio or television signals through his new bilateral hearing aids. He reports he has begun to hear voices when he wears his hearing aids.

47.1 Clinical History and Description

MT, an automotive repair shop owner and mechanic, received his fourth pair of hearing aids 8 months ago. At the fitting, MT stated he was very pleased with his newest bilateral hearing aids and reported that his hearing with these aids has been the best he has experienced when compared with his previous hearing aids worn over the past 20 years. At that point, MT was scheduled for hearing aid maintenance checks every 4 months. At his first 4-month hearing aid maintenance appointment, MT reported dissatisfaction with both hearing aids and believed they were providing less amplification than they had when they were dispensed 4 months earlier. At this appointment, the audiologist cleaned the microphones and receivers in the hearing aids and the resulting electroacoustic analysis revealed MT's hearing aids were performing as expected based upon the previous electroacoustic measures at the time of the dispensing. As he was leaving the clinic, MT expressed gratitude and satisfaction with the clarity of sound his hearing aids were now producing following the maintenance provided during this visit. Four months later, MT returned, reporting that both hearing aids were now receiving radio or television signals because he now hears voices when wearing his hearing aids.

47.2 Questions for the Reader

1. What is significant about MT's occupation as it relates to his most recent report about his hearing aids?
2. What is significant about MT being scheduled for 4-month hearing aid checks?
3. What may be some possible explanations for MT hearing voices while wearing his hearing aids?
4. What tests and services are appropriate in providing the best care for this patient?

47.3 Discussion of Questions to the Reader

1. What is significant about MT's occupation as it relates to his most recent report about his hearing aids?
   It is important to consider the environment in which MT spends the majority of his time. As an owner of an automobile repair facility, MT is probably in and out of excessively hot cars in the summer months. There may also be varying exposure to intermittent dust as well as emissions from cars. In addition, MT may also accumulate a great deal of sweat that is mixed with the dusty particulate matter accumulating on his hearing aids over a shorter period of time than is the case for most hearing aid users.
2. What is significant about MT being scheduled for 4-month hearing aid checks?
   As part of best practice at MT's audiology clinic, MT's hearing aids underwent electroacoustic 2 cc coupler analysis in preparation for the initial dispensing of the hearing aids to MT. This initial analysis for the purposes of quality control assures that upon arrival the electroacoustic performance of MT's hearing aids met the manufacturer's specifications. In addition, and equally important, is that after programming was completed, the 2 cc electroacoustic performance of MT's hearing aids was repeated to serve as a reference for future comparisons. The 4-month post–hearing aid fitting hearing aid check includes the following services:
   a) Clean the microphone ports and replace microphone filters (if present).
   b) Replace the earhooks (if present).
   c) Replace the wax guards and provide the patient with extra packets to last 4 months.
   d) Clean the battery compartment, microphones, and receivers with 99%alcohol then suction all ports.
   e) Dehumidify the hearing aids.
   f) Clean the earmolds and dry the tubing.
   g) Conduct electroacoustic verification with a 2 cc coupler and compare with the 2 cc analysis at the time the hearing aids were dispensed.

   As already mentioned, at the 4-month warranty appointment, both hearing aids were initially not functioning properly according to MT's reports and confirmed by the 2 cc analysis. MT's 4-month warranty appointment happened to occur during the early part of summer when there was an increase in rainfall and temperature (i.e., higher humidity levels). It is especially troublesome when individuals are frequently going from air conditioned environments to the hot and humid outdoors, as MT typically does. Depending upon the environment and the wearer, it is not unusual for sweat to coat the hearing aid body and find its way to many of the internal components. One obvious symptom of sweat causing problems with hearing aids is when both hearing aids malfunction at the same time in a similar manner. Because MT was provided with a desiccant jar at dispensing, he was reminded about the importance of placing both hearing aids in the jar at the end of each day.
3. What may be some possible explanations for MT hearing voices while wearing his hearing aids?
   There are many physical and/or mental conditions the symptoms of which include the phenomenon of auditory hallucination. An auditory hallucination is a significant diagnostic indicator for psychiatric disorders such as schizophrenia or mania, but it can also manifest as a result of intense stress, sleep deprivation, drug use, or high consumption of caffeine. It is also valuable to ascertain whether the voices are present when MT is not wearing his hearing aids or if MT ever

experienced this problem in the past. The following are some simple questions that MT could be asked about the voices:

a) Do the voices only occur while he is wearing the hearing aids?
b) What do the voices say?
c) Are the voices distressing or comforting?
d) Has he heard the voices at any other time in his past?

MT described the voices as being indiscernible, but similar to a radio tuned between two stations and hearing two different stations simultaneously.

Rather than make an immediate suggestion for a referral to his primary care physician and ultimately a psychiatrist for auditory hallucinations, it would be wise for the clinician to first thoroughly investigate the performance of the hearing aids as a source of the symptoms.

4. What tests and services are appropriate to provide the best care for MT for his current complaint?

It is important to remember that an ear of a clinician with normal hearing doing a listening check may not perceive a stimulus in the same manner as an ear having a hearing loss. A listening check by a clinician in attempting to elicit the same symptom as described by a patient can provide valuable information. Unfortunately, the listening check in this case ended as being auditorially very uncomfortable because MT's instruments were amplifying at an unusually high output level.

As part of the hearing aid check in this case, a thorough cursory inspection of each instrument was completed to determine if there was any evidence of rust or trapped moisture. In this case, the inspection revealed an accumulation of moisture and some rust in the battery compartment of both instruments.

Because electroacoustic measures were completed at the time of dispensing as well as during the 4-month hearing aid check, there were valuable reference measures available for comparison during this visit. In this case it became possible to monitor the effect of MT's environment and its impact on the performance of the hearing aids over time.

47.4 Outcome

At this last visit, electroacoustic analysis revealed that both instruments were performing equally, and the results for both hearing aids did not match the performance of previous measures. For simplicity, only results for the right hearing aid are reported because the performance of the left hearing aid was similar. As seen in the sequential electroacoustic measures (when dispensed, 4-month visit, and 8-month visit) of the right hearing aid (Table 47.1), some of the electroacoustic measures characteristics changed significantly from the time the hearing aid was dispensed to when measures were made at 4 and 8 months. At the time the hearing aids were dispensed (first column in Table 47.1); the full-on gain for an input level of 50 dB sound pressure level (SPL) was 44 dB. At 4 months (second column), after MT had worn the hearing aids in a humid environment resulting in the accumulation of sweat and debris, the full-on gain decreased to 41 dB, which resulted in less than optimal performance of the hearing aids. Also notice the decrease in output saturation sound pressure level at 90 dB (OSPL90), average OSPL90, and an increase in the equivalent input noise level. At that visit, after dehumidifying the hearing aids and cleaning the microphones, receivers, and battery compartments, the hearing aids returned to optimal performance, and the subsequent results were consistent with those recorded at the time of dispensing. At the next hearing aid check appointment after MT wore the hearing aids for 4 months in the vacillating environment between air conditioning and heat and humidity, there was evidence of accumulation of moisture on the internal components. As seen in the last column in Table 47.1, at this visit there was a significant difference in coupler response compared with the 2 cc measures obtained at

Table 47.1 Sequential electroacoustic measures of MT's right hearing aid at dispensing, 4 months and 8 months

Right hearing aid	At dispensing	4 months	8 months
Gain control full-on			
OSPL90 (dB SPL)	124 @ 1600 Hz	122 @ 1600 Hz	121 @ 1500 Hz
Average OSPL90 (dB SPL)	118 dB	115 dB	115 dB
Average gain @ 50 dB SPL	44 dB	41 dB	54 dB
Gain control at reference test position			
Average gain @ 60 dB SPL	40 dB	36 dB	52 dB
Frequency range (Hz)	265–7100	335–7100	710–7100
Equivalent input noise	35 dB	39 dB	46 dB
500 Hz distortion (%)	3%	5%	7%
800 Hz distortion	1%	1%	3%
1600 Hz distortion	0%	1%	3%
Frequency attack/release			
250 Hz	10/0 ms	10/0 ms	15/0 ms
500 Hz	155/0 ms	150/0 ms	100/0 ms
1000 Hz	25/120 ms	10/0 ms	10/180 ms
2000 Hz	740/0 ms	650/0 ms	10/0 ms
4000 Hz	10/0 ms	580/0 ms	10/0 ms

Abbreviation: OSPL90, output saturation sound pressure level at 90 dB; SPL, sound pressure level.

the time the hearing aids were dispensed. In fact, the full-on gain increased to 54 dB. Also notice the continued increase in the equivalent input noise level. The increased gain was considered excessive for MT's hearing loss. Once again, after a comprehensive cleaning and reprogramming to the initial settings, MT expressed satisfaction and reported that he no longer heard voices.

When a patient is in an environment such as MT's or if a patient has an endocrinological disorder with resultant excessive sweating, it is not unexpected to find hearing aids behaving as reported by MT. Although MT no longer heard voices, it was apparent that his lifestyle would result in a repetition of the same hearing aid problems. Both of MT's hearing aids were sent to the manufacturer for an internal protective coating shield to keep moisture from damaging and interfering with internal processors and transducers. MT also purchased an electronic dehumidifier where he would store both hearing aids at the end of the day. MT declined sweat sock covers because the internal protective coating, electronic dehumidifier, and quarterly hearing aid cleanings adequately protected his hearing aids from sweat and debris.

47.5 Key Points

1. It is important to offer patients regularly scheduled hearing aid checks that include coupler measures and also allow the clinician to listen to unusual reports from a patient concerning hearing aid performance.
2. When hearing aids are exposed to excessive moisture and debris a wide range of unusual results can occur.
3. Remember that internal protective coating to hearing aids can be added after the initial dispensing.

Suggested Reading

Frye G. Electroacoustic testing of hearing aids and standards. In: Valente M, ed. Hearing Aids: Standards, Options, and Limitations. 2nd ed. New York: Thieme; 2002:1–63

48 You're Wearing the Best Hearing Aids

Jackie L Clark

EW is a 70-year-old first time hearing aid user who is dissatisfied with the hearing aids he recently purchased at another facility. Unfortunately, that facility refused to provide any reprogramming or modifications once the hearing aids were dispensed, and he arrived at another clinic for follow-up care.

48.1 Clinical History and Description

EW recently purchased his first set of hearing aids from another facility and was assured that he received the most "advanced and sophisticated technology" available. Unfortunately, EW reports significant dissatisfaction with the hearing aids. In addition, EW was unable to return to the other facility within the trial period because his wife was in the hospital for 2 months. When EW stated to the clinician at the other facility that he could not hear well when wearing the hearing aids, not only did the clinician refuse to make modifications, but EW was told, "these are the best hearing aids and they are programmed absolutely perfectly." Because EW felt an urgency to clearly hear his soft-spoken wife, EW scheduled a hearing evaluation and hearing aid consultation at another clinic and agreed to pay for the necessary services. EW's goal for the appointment was to obtain help so he could hear better with his current hearing aids or consider purchasing another pair of hearing aids. At the appointment EW brought a copy of the results of the audiometric examination from the other facility. The clinician at this other facility could easily determine that EW's hearing aids were not the most "advanced and sophisticated technology" available.

A review of EW's case history revealed that even though EW is 70 years old, he is in good physical and mental health and reportedly takes no prescription medication. When EW was 30 years of age he was discharged from the Marine Corps where he had been trained as a helicopter maintenance worker. EW had been forced to retire from his job as an aeronautical engineer 6 years earlier when his wife's health began to decline. EW reports that he rarely hunts anymore, but as a younger man, he would hunt or engage in target practice a few times a week. Now, he only shoots his rifle a few times a month, but only recently started wearing hearing protection.

48.2 Audiological Testing

Audiological results from the other facility revealed a bilateral symmetrical moderate to severe sensorineural hearing loss. Word recognition scores under earphones were 82% and 76% for the right and left ears, respectively, with a presentation level at 30 dB sensation level (SL) (re: speech recognition threshold [SRT]) using monitored live voice. These results indicate slight difficulty in the ability to recognize speech. Immittance audiometry was either not completed or the other facility did not document the results (Table 48.1).

48.3 Questions to the Reader

1. What additional information must you obtain before proceeding?
2. What salient issues are important to keep in mind from the case history?
3. What additional information should be asked about EW's hearing aids?
4. What additional tests are required in order to obtain a better understanding of EW's problem?

48.4 Discussion of Questions to the Reader

1. What additional information must you obtain before proceeding?

   It is important to keep in mind that best practice for hearing aid dispensing dictates the necessity of obtaining an audiological examination no more than 6 months before hearing aids are dispensed, medical clearance or waiver of medical clearance, and appropriate verification measures (e.g., real ear measures) reporting that the measured real ear insertion gain (REIG) or real ear aided response (REAR) arrives as close as possible matching a validated prescriptive target (e.g., DSL vol. 5 or NAL-NL2) at the time of dispensing hearing aids.

   As part of the case history, EW reported a history of excessive exposure to noise occupationally and recreationally; thus it is important to identify all sources of noise exposure, both occupational and recreational during his life.

Table 48.1 Audiological results from the other facility

Ear	250 Hz	500 Hz	1000 Hz	2000 Hz	4000 Hz	8000 Hz	WRS %	dB SL
Right—air conduction	45	45	55	60	80	85	82%	30
Right—bone conduction	45	45	45	55	70			
Left—air conduction	55	50	45	65	80	75	76%	30
Left—bone conduction	DNT	DNT	DNT	DNT	DNT			

Abbreviations: DNT, did not test, WRS, word recognition score; SL, sensation level.

Because EW's wife is currently experiencing significant health issues, it is important for the clinician to learn as much as possible about EW's listening environment at home. For example, if his wife is bedridden and there are medical devices, radio, or television creating additional environmental distractions that could interfere with EW's need to hear his wife's soft voice, his clinician must be made aware of this?

Other bits of important information could be to determine whether EW's dexterity is adequate for the hearing aids he is wearing, or does EW have any family members who have hearing loss? Being able to identify the nature, degree, and onset of hearing loss in family members may provide some information about EW's hearing stability over time.

2. What salient issues are important to keep in mind from the case history?
   Though, it is easy to blame the "other facility" when a hearing aid fitting has gone awry, it is more important for a clinician to focus on identifying EW's specific hearing needs and priorities. EW's wife is reportedly soft spoken and has health issues requiring EW to hear and understand her. Also, it is important to be sensitive to the fact that EW has many stress factors occurring at this time, and acclimatization as well as required care and maintenance of his hearing aids can potentially contribute even more to his stress level. Patients experiencing stress typically require written as well as oral instructions with a possible need for repetition.
3. What additional information should be asked about EW's hearing aids?
   It is known that EW reports "nothing but trouble" from his new hearing aids. Consequently, it is critical to determine when the hearing aids are troublesome. The following are some of the questions that EW may need to answer:
   a) Does EW have difficulty hearing in quiet, noise, or both?
   b) Are there specific environmental sounds that are especially unpleasant?
   c) Are soft sounds excessively loud or unpleasant?
   d) How many hours a day has EW been wearing his hearing aids?
   e) Can EW demonstrate successful insertion and removal of his hearing aids?
   f) Can EW demonstrate successful removal and placement of his hearing aid batteries?
   g) Do EW's hearing aids feel comfortable for a long period of time?

   Though little at this point is known about EW's lifestyle, it is important to determine whether specific hearing assistive technology (HAT) could provide additional benefit. There is also a need for EW to quantify his perception of handicap (disability) using a self-assessment of hearing aid benefit and/or satisfaction.
4. What additional tests are required in order to obtain a better understanding of EW's problems?
   Before engaging in any formal audiological examination, an otoscopic inspection is critical to rule out excessive cerumen or debris. Immittance testing with acoustic reflex thresholds (ARTs) complement the behavioral testing to identify the site of auditory dysfunction. Additionally, otoacoustic emissions (OAEs) may be necessary to rule out pathology of the ear. Audiological and speech audiometry must be repeated within 6 months of dispensing hearing aids, and in this case it is especially important to assure EW's hearing status has not changed since he was seen at the other facility. The more recent audiological examination also included the half-octave frequencies that can allow hearing aids to be more accurately programmed. EW understands that repeating the audiometric tests will likely result in his insurance company denying coverage, and EW will be required to pay for the audiometric examination.

   It is important for clinicians to remember that communication occurs in many listening environments, and communication typically does not use monosyllabic words and talking in a quiet listening environment. Also, speech audiometry performed at the other facility unfortunately used monitored live voice rather than the recorded presentation. Another approach to evaluating speech recognition is to use ear-specific sentence in noise testing (e.g., QuickSIN, Etymotic Research, Inc., Elk Grove Village, IL; hearing in noise test [HINT]; or CID-Everyday Sentence Test with Multi-Talker Babble) at a comfortable presentation level (30–40 dB SL, re: SRT), and these tests may provide more valid information for the programming of hearing aids than evaluating word recognition in quiet.

   After visual inspection of the hearing instruments as well as measuring performance via electroacoustic measures it was determined that the hearing aids were performing appropriately. At this point, it is also important to complete real-ear microphone measures for each hearing aid. Real ear measures will determine how the measured REAR or REIG arrives as close as possible to NAL-NL2 for input levels of 50 (soft), 65 (average), and 80 (loud) dB SPL.

48.5 The Evaluation

EW agreed to pay the clinic for repeating the audiological test completed at the other facility plus the additional real ear measures. EW remarked that he did not recall the other facility providing such thorough audiological care nor completing some of the additional measures completed at the second clinic. Otoscopy was unremarkable bilaterally, and immittance audiometry revealed normal tympanic membrane mobility with elevated ARTs bilaterally.

48.6 Additional Case Information

Upon questioning, EW reported that he wore his new hearing aids only a few hours a week and has worn the hearing aids periodically since his wife was admitted to the hospital. EW expressed satisfaction with his hearing aids only when his listening environment was quiet. EW also stated that his hearing aids were comfortable to wear. Visual inspection confirmed that EW could appropriately insert and remove both hearing aids successfully with no apparent challenges. EW's primary goal with his current hearing aids was to better understand his wife as well as to hear better in church. EW's typical listening environment has a mild to moderate amount of background noise. Also, his wife keeps the television or radio on during her waking hours, which reduces the effectiveness of EW's hearing aids. As a means of relaxing, EW enjoys watching the news and some

Table 48.2 Audiological results at the current facility

Ear	250 Hz	500 Hz	1000 Hz	2000 Hz	3000 Hz	4000 Hz	6000 Hz	8000 Hz	SRS + 5 SNR	SRS + 10 SNR	dB SL
Right—air conduction	45	45	55	65	75	80	90	85	0%	10%	40
Right—bone conduction	50	45	45	55	70	70					
Left—air conduction	55	50	45	55	80	80	90	75	32%	64%	40
Left—bone conduction	DNT	DNT	DNT	DNT	DNT	DNT					

Abbreviations: DNT, did not test, SL, sensation level; SNR, signal to noise ratio; SRS, sentence recognition score.

sports programs on his own television while his wife listens to television in her room. Telephone communication is very difficult without his hearing aids, and with his hearing aids, communication on the telephone is even more challenging. Results from the Hearing Handicap Inventory–Elderly (HHIE), a self-assessment tool designed to assess the effects of hearing impairment on the emotional and social adjustment of elderly people, revealed that EW perceives a moderate (score of 40) hearing handicap in the Social and Emotional sections of the HHIE.

48.7 Behavioral Test Results

Pure-tone thresholds using insert earphones were within ± 10 dB of the thresholds measured at the other facility. EW experienced significant difficulties with sentence recognition (presented at dB SL, re: SRT) in + 5 dB signal to noise ratio (SNR) multitalker babble (0% and 32% correct for the right and left ears, respectively) as well as in a + 10 dB SNR (10% and 64% for right and left ears, respectively) when using a 40 dB SL presentation level (Table 48.2).

48.8 Hearing Aid Programming

With the hearing aids connected to the programming software, datalogging confirmed the limited time and the environmental noise levels reported by EW. Though EW's hearing aids have t-coils, they had not been activated and only an omnidirectional with fast (75 ms) release time was selected. Recent research provides evidence that elderly patients with mild cognitive impairment perform better with slow release time. Though EW is not demonstrating behaviors of mild cognitive impairment, his age paired with his hearing complaints and the current stressful situations justified programming his hearing aids with a slower release time (e.g., 800 ms). In addition, to provide greater benefit in noisy listening situations, his hearing aids were reprogrammed with Program 1 using an omnidirectional microphone for quiet listening environments and Program 2 using an adaptive directional microphone for noisy listening environments. The t-coil was activated in both hearing aids as well.

48.9 Hearing Instrument Results

Both hearing aids were cleaned and electroacoustic measures revealed they were performing according to manufacturer specifications. Unfortunately, EW's wife, due to her physical and cognitive limitations, was unable to attend the appointment to provide her voice for real ear measures. Speech mapping real ear measures were completed using a speech-weighted signal presented at 55, 60, and 75 dB sound pressure level (SPL). The measured REAR matched the National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) prescriptive targets and the SSPL90 was below the uncomfortable loudness (UCL) target.

Directional microphone performance was verified using the front-to-side (FSR) ratio by calculating dB SPL output in response to stimuli presented from 0 to 360 degrees using the hearing aid analyzer chamber. The responses of the t-coils were verified using the Telephone Magnetic Field Simulator (TMFS) to measure the Relative Simulated Equivalent Telephone Sensitivity (RSETS). This measure confirmed an RSETS of 0 dB, indicating that the frequency response of the telecoil (T-coil) matched the frequency response of the microphone.

In order to verify the effectiveness of each hearing aid program, EW's aided performance for sentence recognition in noise was completed. While wearing the recently programmed hearing aids bilaterally, EW was seated in a sound suite at 90° and 1 meter from a loudspeaker. Recorded sentences using a male talker at 60 dB HL were simultaneously presented with a four-talker multitalker babble presented at 55 dB HL (+ 5 dB SNR) from the same loudspeaker. With the hearing aids in Program 1, EW achieved a score of 40% With the hearing aids in Program 2, EW's score improved to 60%

48.10 Outcome

EW was counseled on the changes made to his hearing aids. EW demonstrated success in communicating during telephone conversation after he switched to the t-coil program. There was some discussion concerning the difference between "hearing" and "understanding" speech. Once this was explained, EW understood the benefit of hearing aids to amplify signals as well as recognizing the limitations of hearing aids to clarify speech signals. Though EW initially indicated a readiness to purchase new hearing aids, he was willing to defer any decision concerning purchasing new hearing aids until he wore his current hearing aids with the recent programming for 30 days. When it was explained to EW that his hearing aids would continue datalogging his listening time and environment, EW agreed to be more vigilant in wearing his hearing aids for at least 8 to 10 hours daily. EW called 30 days after the appointment to report satisfaction with his current hearing aids, and he believed there was no need to replace the hearing aids.

48.11 Key Points

1. This case provides evidence about the importance of a comprehensive case history along with a thorough diagnostic examination that includes measures of speech in noise performance.
2. Oftentimes, speech in noise performances will not only provide better diagnostic information but will also objectively measure success of a remediation plan and outcome of the hearing aid fit.
3. This cases shows importance of including the t-coil in hearing aids so patients can improve communication on the telephone.
4. This cases shows importance of using real ear measures to verify the performance of hearing aids.
5. This cases shows importance of charging patients for the services provided in one clinic for hearing aids purchased at another clinic or in another manner (online)

Suggested Reading

Putterman DB, Valente M. Difference between the default telecoil (t-coil) and programmed microphone frequency response in behind-the-ear (BTE) hearing aids. J Am Acad Audiol 2012; 23: 366–378

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Ventry IM, Weinstein BE. The hearing handicap inventory for the elderly: a new tool. Ear Hear 1982; 3: 128–134

Wu YH, Bentler RA. Clinical measures of hearing aid directivity: assumption, accuracy, and reliability. Ear Hear 2012; 33: 44–56

E-GIRQ.COM

49 Liquefaction (Hydrolytic) Necrosis from a Hearing Aid Battery in the Ear Canal (Assault and Battery in the Ear Canal)

Cynthia Collier and Ross J. Roeser

In this case report, SR is an 84-year-old woman presenting with a no. 312 hearing aid battery that had been in her right ear canal for several weeks. This is a situation that can lead to liquefaction (hydrolytic) necrosis.

49.1 Clinical History and Description

SR was an 84-year-old woman who was first seen for a comprehensive audiological examination in January 2010. She returned 3 months later indicating that her ability to recognize speech had decreased.

49.2 Audiological Testing

SR's initial audiological examination was in January 2010 and her primary complaint was an inability to recognize conversations in most listening situations. Pure-tone audiometry revealed a bilateral symmetrical mild to severe sensorineural hearing loss that is gradually sloping in configuration. Speech recognition thresholds (SRTs) revealed a moderate loss in the ability to receive speech bilaterally. Word recognition scores (WRSs) using full-list recordings of the Northwestern University Auditory Test No. 6 (NU-6) word lists revealed a slight loss in the ability to receive speech bilaterally. Tympanograms revealed normal ear canal volume and static compliance, as well as normal middle ear pressure. Contralateral and ipsilateral acoustic reflex thresholds (ARTs) at 1000 Hz were normal (Fig. 49.1). SR's responses to the Hearing Handicap Inventory–Elderly indicated a mild-to-moderate handicap. QuickSIN testing (Etymotic Research, Inc., Elk Grove Village, IL) presented at 70 dB HL revealed a moderate signal to noise ratio (SNR) loss, bilaterally (12 dB).

SR was fitted bilaterally with digitally programmable receiver-in-the-ear (RITE) instruments that used no. 312 batteries. The fitting required more counseling than usual, due to the patient's reduced understanding of use and care of the instruments. The patient, however, presented with no appearance of cognitive decline. SR returned to the clinic for a routine hearing aid check in mid-February, at which time electroacoustic analysis revealed proper functioning of the hearing instruments.

Three months later, in May 2010, SR returned for a follow-up visit with a report that she was receiving reduced benefit from her instruments. Otoscopy revealed debris in the right ear canal close to the tympanic membrane. SR denied any perception of this obstruction. The debris appeared to be dark cerumen, but when removal was attempted with an ear curette it was found to be solid, hard, and firmly adhering to the canal wall. Removal of the surface debris revealed a shiny silver object, which was identified as a no. 312 hearing aid battery.

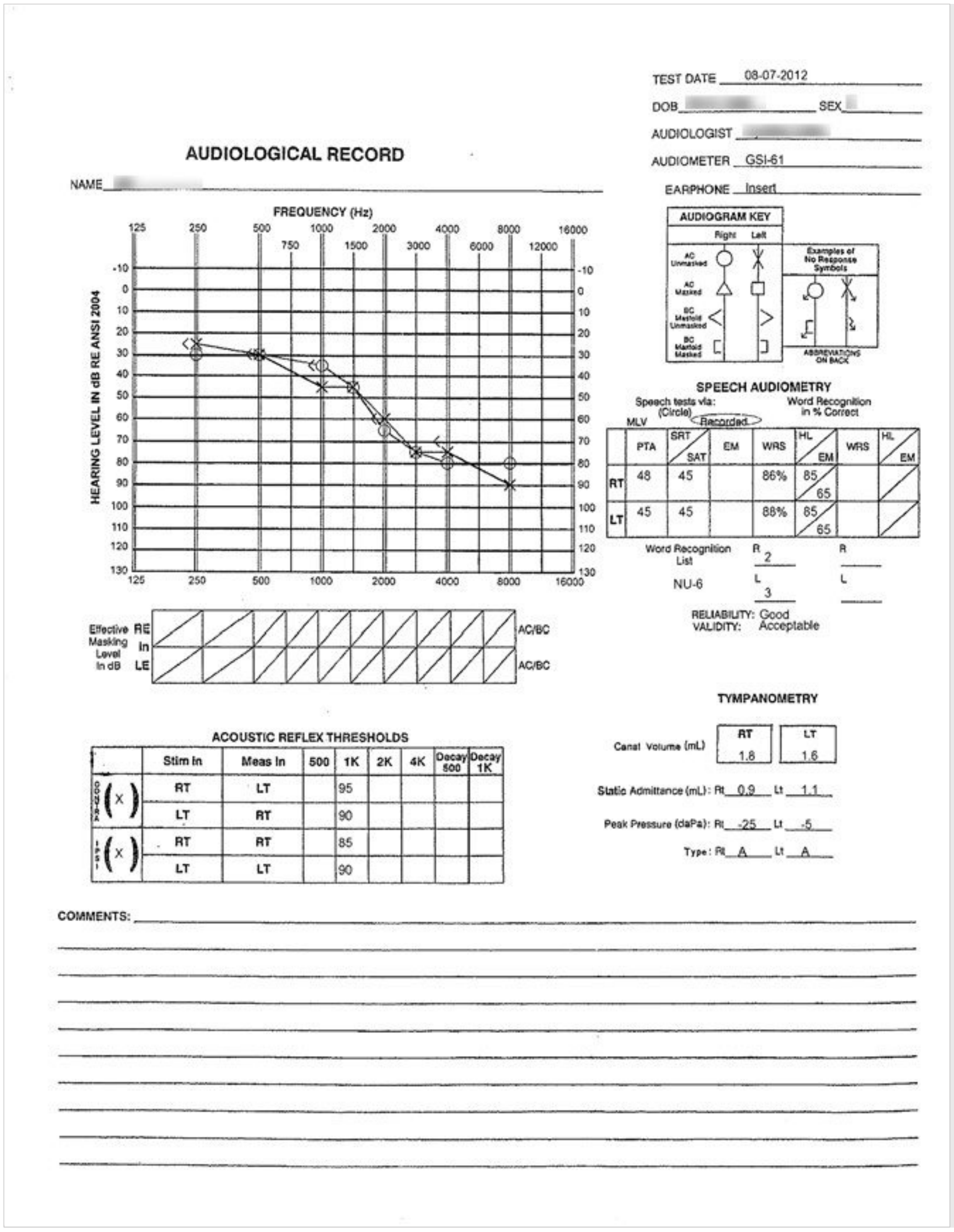
49.3 Questions to the Reader

1. What factors must be considered prior to attempts to remove the battery from the ear canal?
2. What safe and effective removal procedure(s) should be considered?
3. If attempts are unsuccessful, or if removal is not attempted, what referral(s) should be made?
4. What potential consequences exist for a battery lodged in the ear canal?

49.4 Discussion of Questions to the Reader

1. What factors must be considered prior to attempts to remove the battery from the ear canal?
   Removing any object from a patient's ear canal, particularly a hearing aid battery, as soon as possible is always important. Removal in a nonmedical facility is possible if the object (hearing aid battery) is not adhering to the ear canal wall and can be removed without force. If, however, the patient is on blood thinners, is immunocompromised, or is diabetic, medical removal is advised because even a small scratch of the ear canal can result in negative outcomes in these patients. Further, if the object is stubbornly adhering to the canal wall, otologic referral is indicated. This is particularly the case when the object is identified as a hearing aid battery embedded in cerumen. In fact, such cases should be considered an audiological/otologic emergency.

   Cerumen has a high electrical conductivity and therefore a low-voltage electrical current can be created in the ear canal when a battery comes into contact with cerumen. Left unchecked, this low-voltage current can result in the exudation of tissue fluids. The resulting moist environment of the ear canal created by these fluids can lead to the spontaneous leakage of electrolytes from the battery, which will generate hydroxide. Hydroxide is a corrosive substance, and its presence in the canal will result in liquefaction necrosis, which can also be referred to as "hydrolytic" necrosis (a term the authors suggest).
2. What safe and effective removal procedure(s) should be considered?
   Several extraction methods might be considered, provided the battery is no adhering to the canal wall: (1) Suction is a safe removal option and is effective; (2) alligator forceps or a right-angle hook can be used to grasp or capture the battery; (3) cyanoacrylic glue can be applied to the blunt wood end of a cotton swab and be held to the battery for 10 seconds in order to allow a bond to form. When the wood probe is removed from the canal, the adhered battery should follow.



FREQUENCY (Hz)
125 250 500 750 1000 1500 2000 3000 4000 6000 8000 12000 16000
HEARING LEVEL IN dB RE ANSI 2004
-10 0 10 20 30 40 50 60 70 80 90 100 110 120 130
AUDIOGRAM KEY
Right Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
ABBREVIATIONS ON BACK

AUDIOLOGICAL RECORD

NAME [redacted]

TEST DATE 08-07-2012

DOB [redacted] SEX [redacted]

AUDIOLOGIST [redacted]

AUDIOMETER GSI-61

EARPHONE Insert

Effective Masking Level in dB RE AC/BC

In LE AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT	48	45		86%	85 / 65		
LT	45	45		88%	85 / 65		

Word Recognition List R 2 R

NU-6 L 3 L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.8	1.6

Static Admittance (mL): Rt 0.9 Lt 1.1

Peak Pressure (daPa): Rt -25 Lt -5

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT		95				
	LT	RT		90				
IPSI (X)	RT	RT		85				
	LT	LT		90				

COMMENTS:

Fig. 49.1 Audiometric findings obtained from SR prior to discovering a no. 312 battery in her right ear canal.

Aural irrigation, by contrast, is absolutely contraindicated in such cases because the water can cause spontaneous leakage of electrolytes. For the same reason, any procedure that might cause excessive bleeding is contraindicated.

3. If extraction attempts are unsuccessful, or if removal is not attempted, what referral(s) should be made?
   As in the case of any obstruction in the ear canal that cannot be successfully removed in an audiology office, referral should be made to a physician, preferably an otologist or otolaryngologist. Unique with regard to foreign objects in the ear canal, a battery in the ear canal should be treated as an emergency. The audiologist or patient should make an immediate appointment with his or her physician as soon as possible or, alternatively, seek treatment in an emergency room.
4. What potential consequences exist for a battery lodged in the ear canal?
   If the battery is not quickly removed from the ear canal there are several possible consequences, progressing medially from the site of the lodged battery: (1) liquefaction necrosis of the canal wall and surrounding tissues, (2) facial paralysis due to caustic injury of the seventh cranial nerve, (3) erosion of the tympanic membrane, (4) ossicular erosion, (5) erosion of the medial wall of the middle ear, and, ultimately, (6) sensorineural hearing loss and damage to the vestibular labyrinth.

49.5 Additional Question to the Reader

1. Are there published reports of otologic damage from a hearing aid battery?

49.6 Discussion of Additional Question to the Reader

1. Are there published reports of otologic damage from a hearing aid battery?
   Many patients who have experienced a button battery in their ear canal(s) have not been as fortunate as SF. For example, Premachandra and McRae described a 10-year-old boy who presented with a button battery that he admitted to inserting into his ear canal 3 weeks earlier. The patient

reported severe otalgia, and otoscopy revealed black otorrhea and gross meatal edema. The corroded button battery was removed under general anesthesia.

Premachandra and McRae also described a 76-year-old woman with dementia who presented with a 3-day history of severe otalgia. Upon removal of a corroded zinc alkaline battery under general anesthesia, necrotic deep meatal skin was observed with underlying bony erosion.

49.7 Diagnosis and Recommended Treatment

Following a failed attempt at mechanical removal of the battery, SR was referred to an otologist for an emergency procedure. The battery was successfully removed with no complications.

49.8 Outcome

Because the battery compartment of the RITE instrument worn by SR is remote from the ear canal (behind the pinna) when it is worn properly, how the no. 312 battery found its way into SR's right ear canal remains a mystery. SR was surprised to learn of its presence and had no recollection of losing a battery. SR was fortunate not to have any observable changes to her right ear canal. Audiological examination following the battery extraction revealed no change in pure-tone thresholds. The patient was reinstructed on the proper care and use of the instruments, especially in regard to proper battery insertion.

49.9 Key Points

1. A battery lodged in the ear canal is an otologic emergency.
2. Mechanical methods of removal can be attempted in the audiologist's office, including suction, glue, a magnet, or alligator forceps if the battery is not adhering to the ear canal wall.
3. Irrigation or any other procedure that would introduce moisture into the ear canal is contraindicated because the use of water can promote electrolyte leakage from the battery.

Suggested Reading

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Dance D, Riley M, Ludemann JP. Removal of ear canal foreign bodies: what can go wrong and when to refer. BCMJ 2009; 51: 20–24

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50 Evaluation of a Fully Implantable Hearing Device

Anna C. S. Kam and Lena L. N. Wong

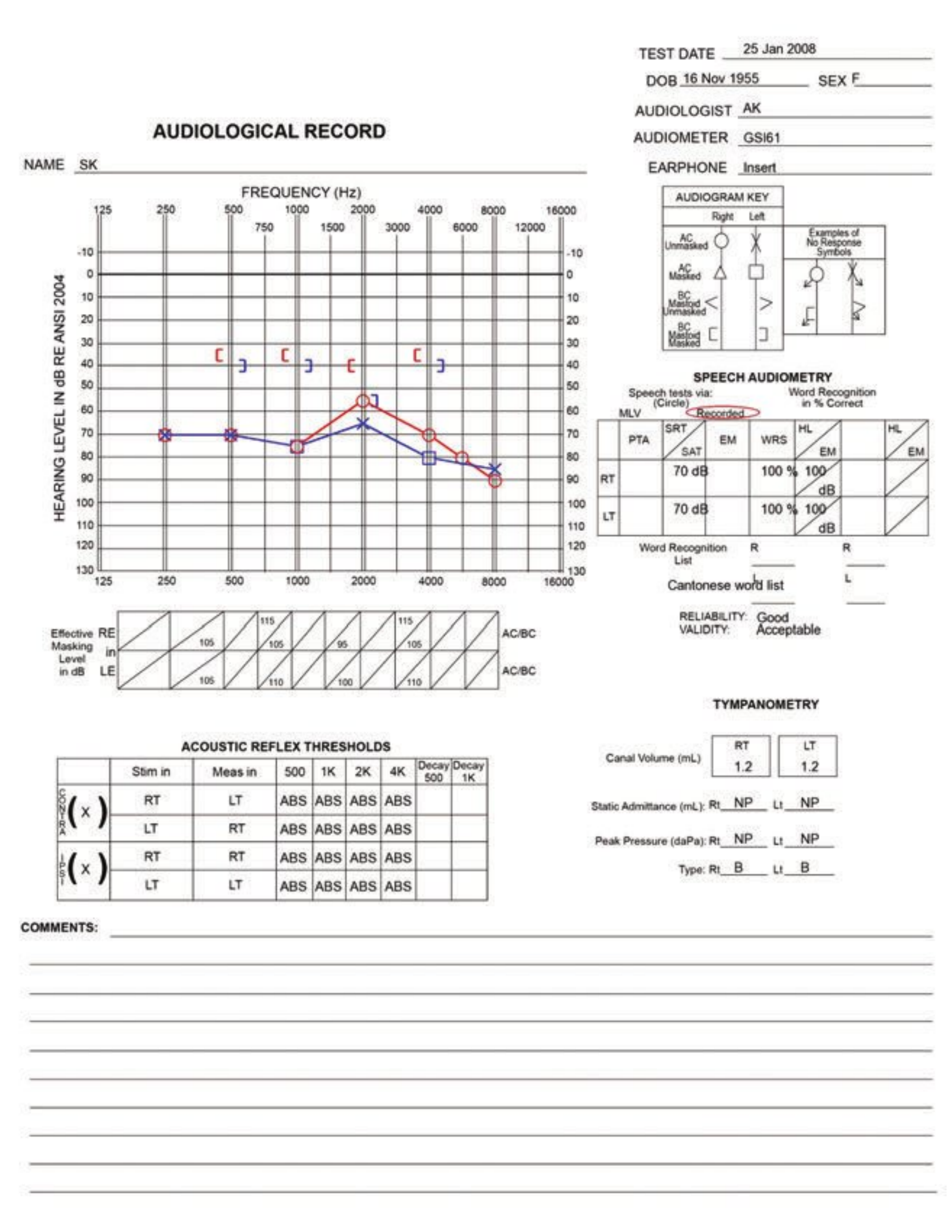
SK is a 57-year-old female with a fully implantable hearing device (FIHD) in one ear and a conventional hearing aid in the opposite ear after many years of unsuccessful use with bilateral conventional hearing aids.

50.1 Clinical History and Description

SK is a 57-year-old female with a bilateral moderately severe to severe mixed hearing loss. An initial diagnosis of otosclerosis was made and an unsuccessful stapedectomy was performed when SK was 33 years of age. A Widex ES-6 H behind-the-ear (BTE) hearing aid (Widex USA, Inc., Hauppauge, NY) was fit to the right ear at that time. For cosmetic reasons, bilateral Phonak 9000 AFSC 312 in-the-canal (ITC) hearing aids (Phonak, Warrenville, IL) were fit 10 years later. With deteriorating hearing, SK was fit with bilateral Beltone M75 BTE hearing aids (Beltone, Glenview, IL) in 2007, but she repeatedly reported concerns related to the appearance and inconvenience of hearing aid use and inquired about an FIHD.

50.2 Audiological Testing

SK's most recent hearing thresholds were found to be consistent with a bilateral moderately severe to severe mixed hearing loss (□ Fig. 50.1) with significant air–bone gaps at 500 to 4000 Hz and decreased bone conduction thresholds. The speech recognition threshold (SRT) revealed a bilateral moderately severe loss in the ability to receive speech and was in agreement with the pure-tone average (PTA). Word recognition scores (WRS) of 100% were revealed bilaterally at a presentation level of 100 dB HL, indicating normal ability to recognize speech at a high input level. Type B tympanograms were



AUDIOLOGICAL RECORD

NAME SK

TEST DATE 25 Jan 2008
DOB 16 Nov 1955 SEX F
AUDIOLOGIST AK
AUDIOMETER GSI61
EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		70 dB		100 %	100 dB		
LT		70 dB		100 %	100 dB		

Word Recognition List R L
Cantonese word list

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	ABS	ABS	ABS	ABS		
	LT	RT	ABS	ABS	ABS	ABS		
IPSI (X)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.2

Static Admittance (mL): Rt NP Lt NP
Peak Pressure (daPa): Rt NP Lt NP
Type: Rt B Lt B

COMMENTS:

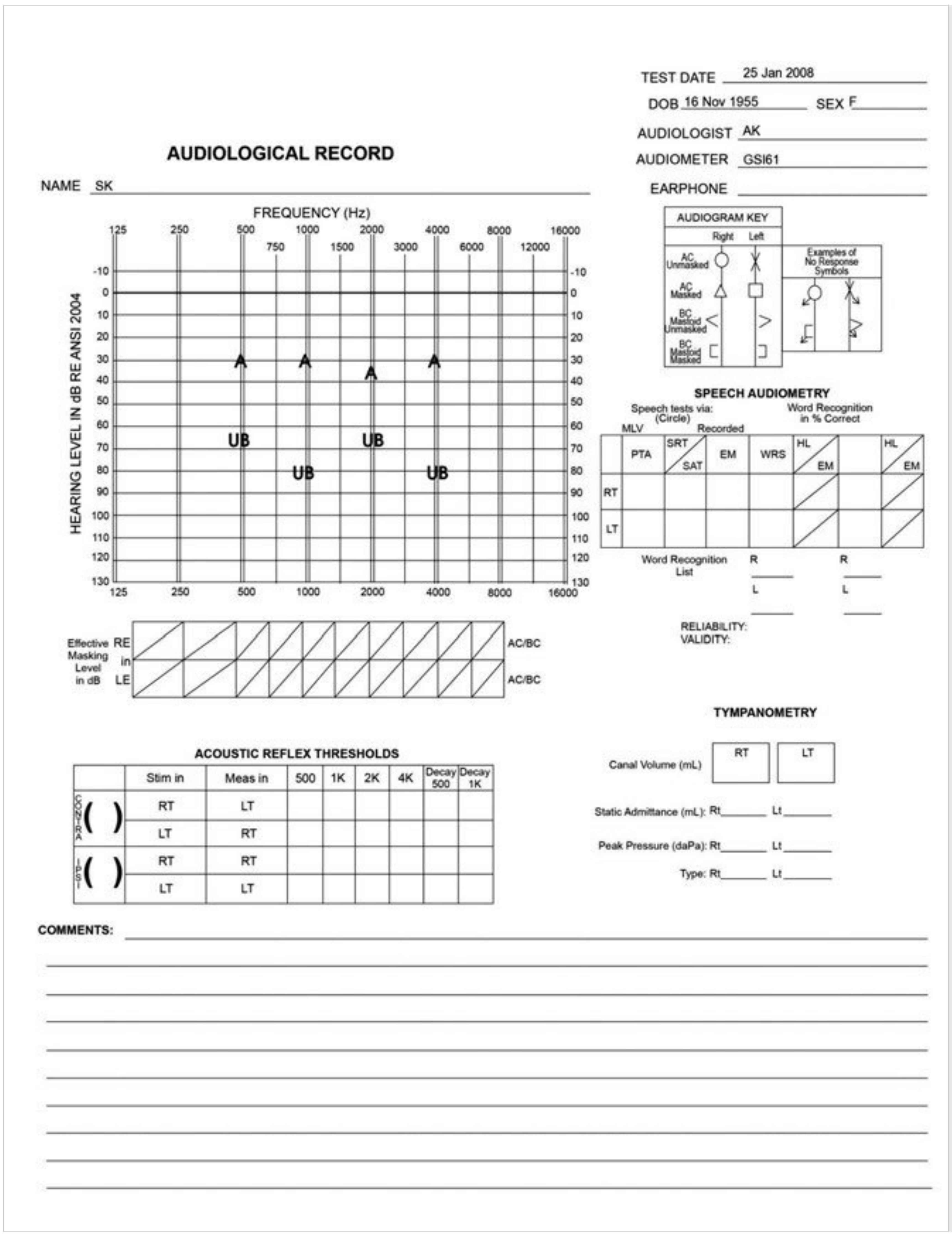
Fig. 50.1 Audiometric results before implanting the fully implantable hearing device (FIHD).

recorded bilaterally with ipsilateral and contralateral acoustic reflex thresholds (ARTs) absent at 500 to 4000 Hz bilaterally. These results support the presence of a conductive component to SK's hearing loss.

Bilateral unaided (UB) and aided (A) sound-field thresholds (dB HL) with the M75 hearing aids were obtained using warble tones at 500 to 4000 Hz in octave intervals with a loudspeaker at 0° azimuth and placed at 1 meter (Fig. 50.2). These results reveal functional gain of 30 to 50 dB depending upon test frequency.

SRTs were measured in quiet (dB HL) and noise (signal to noise ratio [SNR] in dB) for unaided and two aided conditions (bilateral M75; Otologics Carina FIHD (Cochlear Boulder LLC, Boulder, CO) in right ear and M75 in the left ear) using the recorded version of the Cantonese Hearing in Noise Test (CHINT). A conventional adaptive procedure was used where, for the noise condition, the speech-shaped noise was fixed at 65 dB(A) and the presentation level of the CHINT sentences are adaptively adjusted based on SK's response. That is, the presentation levels for the sentences were increased in 2 dB steps for an incorrect response and decreased in 2 dB steps for a correct response to obtain the level where SK could correctly repeat the sentences 50% of the time. For the SRT procedure for the quiet condition, the sentences were presented in isolation. Also, a speech recognition score (SRS in %) was measured using the CHINT sentences at a fixed signal and noise level and again measured for quiet and noise conditions. That is, in quiet, the CHINT sentences were presented at 65 dB(A). For the noise condition, the CHINT sentences were fixed at 70 dB(A) and the CHINT noise was presented at 65 dB(A), yielding an SNR of +5 dB with the CHINT sentences and CHINT noise presented from a loudspeaker at 1 meter at 0° azimuth.

As reported in Table 50.1, unaided SRTs could not be measured using the conventional adaptive procedure or the fixed SNR procedure and the SRSs were 0% in both quiet and noise. At fixed input level in the quiet condition, the aided SRS was 98% with the bilateral M75 hearing aids, and the same score was achieved with the FIHD implanted into the right ear and the M75 fit to the left ear. This finding indicated no difference in performance between the two methods of amplification. At fixed signal level in the noise-front condition, the aided SRS was 82.6% with the bilateral M75 hearing aids and 84.5% was achieved with the FIHD implanted into the right ear and the M75 fit to the left ear. Again, this finding indicated no difference



AUDIOLOGICAL RECORD

TEST DATE 25 Jan 2008

DOB 16 Nov 1955 SEX F

AUDIOLOGIST AK

AUDIOMETER GSI61

EARPHONE

NAME SK

COMMENTS:

Fig. 50.2 Unaided bilateral (UB) and aided (A) sound-field thresholds before implanting the fully implantable hearing device (FIHD). Aided thresholds were measured with the M75 behind-the-ear hearing aids (Beltone).

in performance between the two methods of amplification. Finally, aided SRTs with the bilateral M75's were 50.3 dB HL in quiet and a 2.0 dB SNR in noise, whereas the aided SRTs for the FIHD implanted into the right ear and the M75 fit to the left ear were 51.8 dB HL and 1.7 dB SNR, respectively. Again, these findings report no difference in performance between the two methods of amplification.

Validation measures were obtained using the Chinese version of the Abbreviated Profile of Hearing Aid Benefit (APHAB) and the Satisfaction with Amplification in Daily Life (SADL). The Global Score of the APHAB was 37 (with a score of 100 indicating the greatest benefit) and a satisfaction rating from the SADL was 3.75 (with a score of 7 indicating the greatest satisfaction). SK also reported "fair" sound quality, occasional feedback, and some cosmetic concerns.

50.3 Questions to the Reader

1. Did the hearing aids provide optimal outcome?
2. Is SK a candidate for an FIHD?

50.4 Discussion of Questions to the Reader

1. Did the hearing aids provide optimal outcomes?
   SK's M75 hearing aids were fit using National Acoutic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) corrected for the magnitude of the air–bone gap (75% of the air–bone gap added to the prescriptive target). Real ear measurement (REM) with digital speech signals presented at input levels of 50, 65, and 80 dB sound pressure level (SPL) revealed a maximum of < 6 dB deviation from the NAL-NL1 real ear aided response (REAR) targets for input levels at 50, 65, and 80 dB SPL at 250 to 4000 Hz. SK's aided performance on the SRT and SRS improved when compared with unaided performance (Table 50.1). These results also indicated that SK was able to recognize conversational speech. Self-reported perceived benefit, however, for the M75 hearing aids was only "moderate" and SK was clearly dissatisfied with her M75 hearing aids. Most importantly, SK had cosmetic concerns even though she was using the hearing aids for 8 to 9 hours per day at work.
2. Is SK a candidate for a FIHD?
   Revision middle ear surgery had been explored with SK, but this option was considered inappropriate due to scarring around the incus. Hearing aid trials with more recent technologies were recommended, but SK declined because of cosmetic concerns. SK was informed of the advantages of an FIHD as well as limited evidence related to the use of the FIHD in patients with mixed hearing loss. Possible risks and limitations were also discussed. These include risks related to the use of general anesthesia, extraction of a faulty device, or extraction due to insufficient power when further deterioration of hearing occurs. SK was also informed that she may have to sleep in different positions to avoid applying pressure on the implant. In fact, when SK was first considered for an FIHD, the FIHD was still under clinical trial for sensorineural hearing loss and, to date, the U.S. Food and Drug Administration (FDA) has not approved the FIHD for clinical use. Placement of the transducer on the round window membrane was necessary because of scarring on the incus, thus increasing the uncertainties in the mass loading of the ossicles and its effect on sound transmission to the inner ear. Otherwise, SK's magnitude of hearing loss was within the recommended fitting range (moderate to severe). After thorough informative counseling and medical clearance, SK decided to go ahead with the surgery to fit the FIHD.
   Medical examination including magnetic resonance imaging (MRI) and computed tomographic (CT) scan did not reveal any medical contraindications for the FIHD. An FIHD (Otologics Carina) was implanted on the right side 4 years ago (Fig. 50.3). Continued use of the M75 hearing aid on the left ear reportedly helped SK with improving speech recognition in noise and localization.

50.5 Additional Testing

Previous research reported that, immediately after the FIHD was activated, functional gain ranged from 19 to 36 dB. This initial outcome, however, decreased significantly after 1 year postimplantation. Thus the authors of this case report decided to report the outcomes assessed for SK with the FIHD at 1 year postactivation. No significant change in the unaided hearing thresholds was noted (Fig. 50.4 compared with Fig. 50.1 and Fig. 50.5 compared with Fig. 50.2). Again, a significant improvement in aided sound-field thresholds was found compared with unaided thresholds (Fig. 50.5). Fig. 50.5 reports that aided thresholds did not improve below 30 dB HL at 500 to 4000 Hz, as expected, because of noise floor of the microphone of the FIHD. Aided SRS of 98.0% in quiet and 84.5% in noise with stimuli presented at a fixed level were obtained. Aided SRTs were 51.8 dB HL and 1.7 dB SNR in the quiet and noise conditions, respectively. The SADL Global Score increased from 3.75 to 5.25 (i.e., from slightly > the 20th percentile to > the 80th

Table 50.1 Real ear insertion gain (REIG) measures for input levels of 50, 65, and 80 dB SPL using a DigiSpeech signal for the right (a) and left (b) ear. Also included is the NAL-NL1 target corrected for binaural and channel summation.

Unaided				Aided with bilateral M75^{a} (preoperation)				Aided with Carina FIHD^{b} on the right ear and M75 on the left ear (postoperation)			
Adaptive		Fixed		Adaptive		Fixed		Adaptive		Fixed	
(dB HL)	(SNR)	(% correct)		(dB HL)	(SNR)	(% correct)		(dB HL)	(SNR)	(% correct)	
Quiet	Noise	Quiet	Noise	Quiet	Noise	Quiet	Noise	Quiet	Noise	Quiet	Noise
CNT	CNT	0	0	50.3	2.0	98.0	82.6	51.8	1.7	98.0	84.5

Abbreviations: CNT, could not test; FIHD, fully implantable hearing device; SNR, signal to noise ratio. ^{a}Beltone, Glenview, IL. ^{b} Cochlear Boulder LLC, Boulder, CO

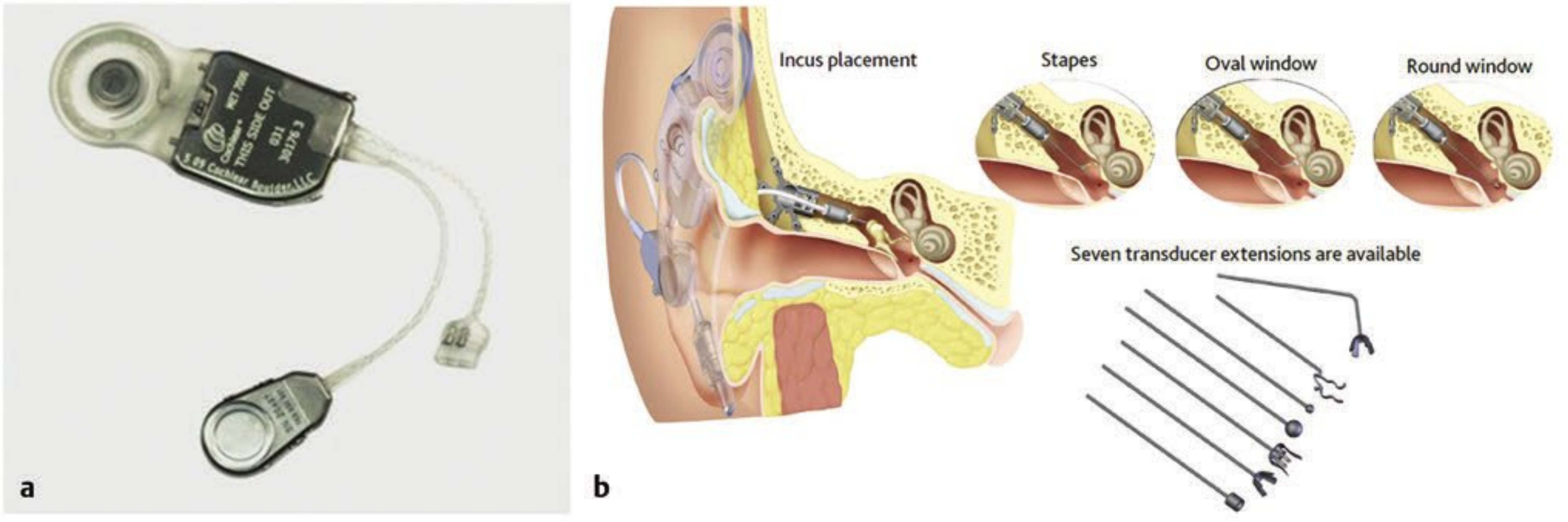
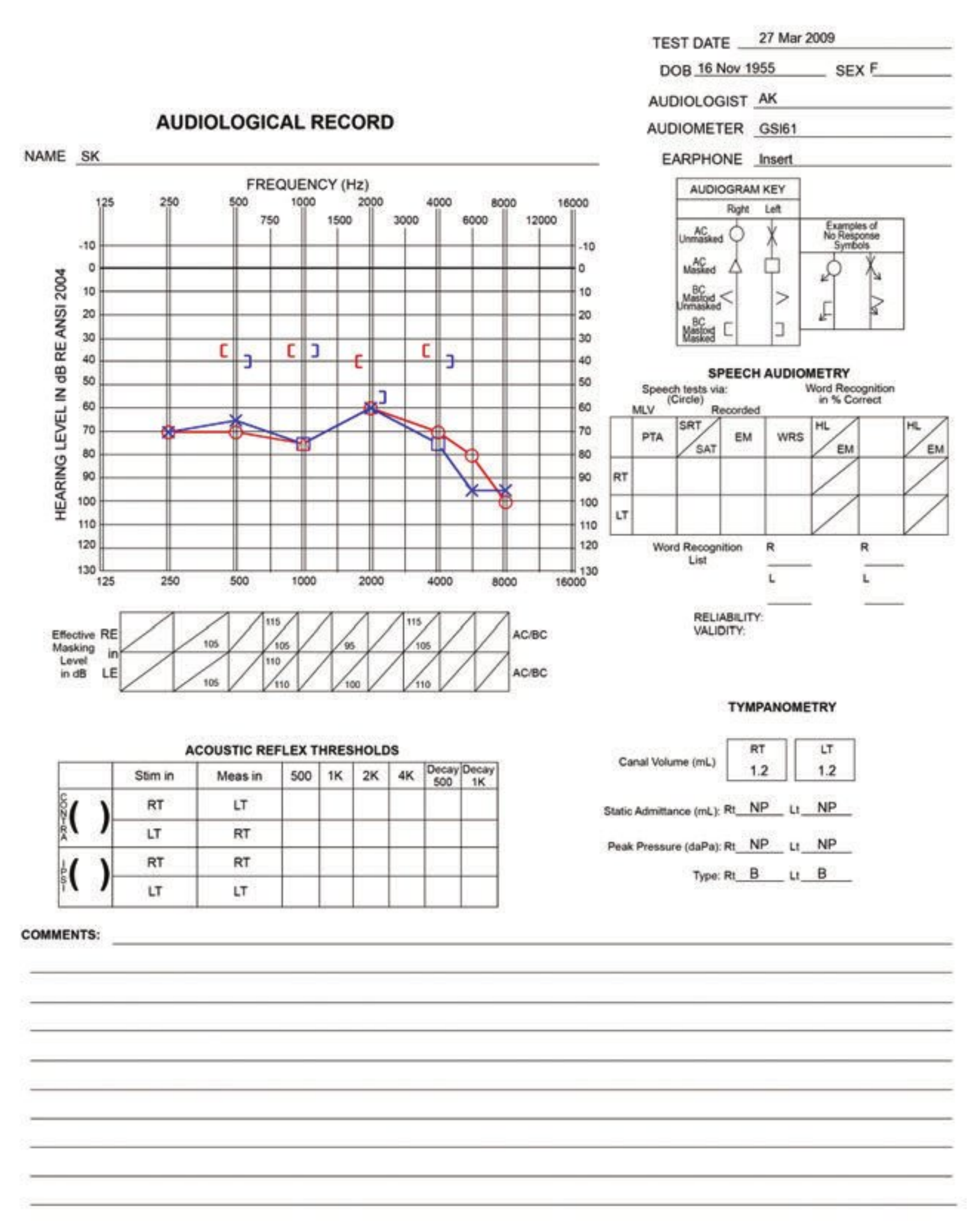


Fig. 50.3 The Carina fully implantable hearing device (a) and the implanted view (b). (Reprinted with permission from Cochlear Boulder LLC.)



TEST DATE 27 Mar 2009
DOB 16 Nov 1955 SEX F
AUDIOLOGIST AK
AUDIOMETER GSI61
EARPHONE Insert

AUDIOLOGICAL RECORD

NAME SK

Effective Masking Level in dB

RE in		105		115 105		95		115 105				AC/BC
LE		105		110 110		100		110				AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT							
LT							

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.2	1.2

Static Admittance (mL): Rt NP Lt NP
Peak Pressure (daPa): Rt NP Lt NP
Type: Rt B Lt B

COMMENTS:

Fig. 50.4 Audiometric results obtained 1 year after implanting the fully implantable hearing device (FIHD) on the right ear.

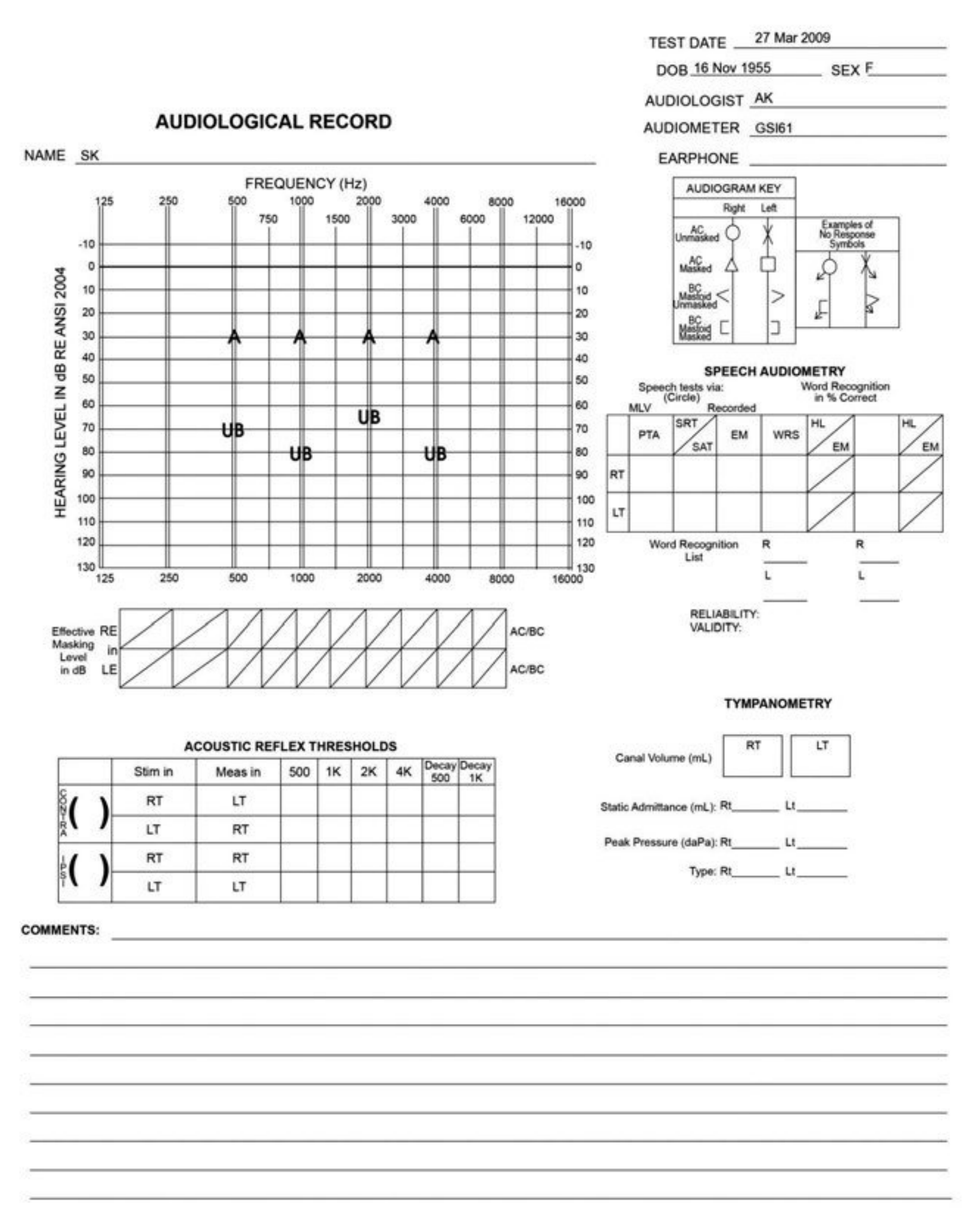
percentile of the normative data). The APHAB Global Rating increased from a score of 37 to 80 (i.e., from >the 50th percentile to >the 95th percentile of the norm scores). Unlike the results reported for the M75, these results suggest a high level of self-perceived aided benefit and satisfaction. These results are commensurate with findings reported in a systematic review on the Carina. These scores also represented a substantial improvement compared with the M75.

50.6 Additional Questions to the Reader

1. Are there other technologies that could have helped SK?
2. Does SK appear to perform better with the FIHD?
3. How was evidence-based practice (EBP) applied in this case?

50.7 Additional Discussion of Questions to the Reader

1. Are there any other technologies that could have helped SK?
   SK primarily complained about the sound quality and appearance of her M75 hearing aids. Though an appropriate fitting of the M75 s was verified using REMs and speech recognition tests, further fine-tuning of the M75 s was attempted but failed to enhance self-reported sound quality. It is possible that hearing aids with more advanced features may have provided improved sound quality. Commercially available miniaturized hearing aids, although suitable for SK's hearing loss, were not sufficient to fully alleviate SK's cosmetic concerns.
2. Does SK appear to perform better with the FIHD?
   Aided sound-field thresholds (dB HL) and speech recognition performance with the FIHD were comparable with the performance obtained with the M75 hearing aids (Fig. 50.1 and Fig. 50.5; Table 50.1). Higher levels of self-perceived aided benefit and satisfaction with the FIHD were noted, however, compared with SK's M75 hearing aids (Fig. 50.6). SK also reported better sound quality with the FIHD and was very satisfied with its performance. Thus, although the FIHD did not outperform the M75 s in the objective measures, the FIHD did provide greater self-perceived aided benefit and satisfaction. It should, however, be noted that, because SK was fully aware of the trial of a new technology and that it is cosmetically more appealing, the report on improved sound



TEST DATE 27 Mar 2009
DOB 16 Nov 1955 SEX F
AUDIOLOGIST AK
AUDIOMETER GSI61
EARPHONE

AUDIOLOGICAL RECORD

NAME SK

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT							
LT							

Word Recognition List R ____ L ____ R ____ L ____

RELIABILITY:
VALIDITY:

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt______ Lt______

Peak Pressure (daPa): Rt______ Lt______

Type: Rt______ Lt______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 50.5 Unaided bilateral (UB) and aided (A) sound-field thresholds obtained 1 year after implanting the fully implantable hearing device (FIHD) on the right ear. Aided thresholds (A) were measured with the FIHD on the right ear and an M75 hearing aid (Beltone) on the left ear.

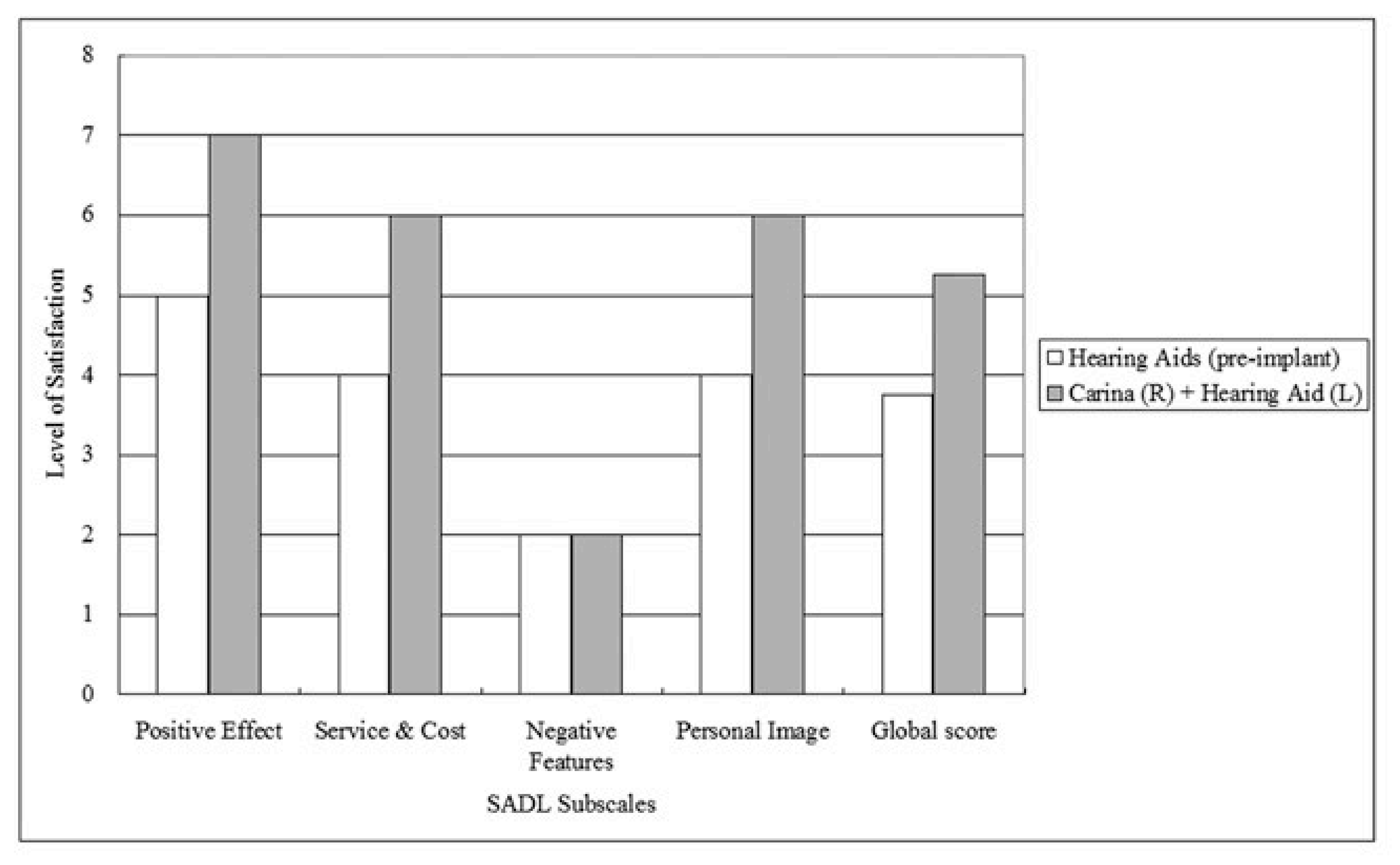


Fig. 50.6 Comparing Satisfaction with Amplification in Daily Life (SADL) ratings obtained using the bilateral M75 hearing aids (Beltone) (preimplant) and postimplant using a fully implantable hearing device (FIHD) (Carina) on the right ear and an M75 hearing aid on the left ear.

quality, benefit, and satisfaction may be a result of a Hawthorne or placebo effect.

3. How was evidence-based practice (EBP) applied in this case?

   Often, clinicians are "forced" to fit hearing devices before best-evidence is available. In EBP, clinicians should consider a patient's preferences and goals as well as the available evidence and appropriate risks that accompany an intervention. Whenever possible, the highest level of evidence (e.g., randomized controlled trial) should be used. In this case, emerging evidence (i.e., lower-level evidence) was available for patients with a similar degree of hearing loss, but there was a lack of evidence for patients with mixed hearing loss. The low-level evidence studies were mainly clinical trials sponsored by the manufacturers of FIHDs and completed with small sample sizes. Outcome measures may be selected by the manufacturers to maximally demonstrate the benefits with these devices that may not be representative of the efficacy in real-life situations. The presence of a Hawthorne effect could not be ruled out. Thus clinicians should be aware of the pitfalls of low-level evidence when recommending an FIHD to patients.

   SK was counseled so that she was fully aware of the advantages as well as possible complications. As evidence accumulates, and with FDA approval in the future, a clinician should feel more confident about recommending, or not recommending, a new intervention such as the FIHD. To date, a newly published systematic review based on research studies with small sample sizes suggests positive outcomes with FIHDs. The analysis also reports an overall failure or malfunction rate of 17.6%because the FIHD did not charge correctly or extruded. In some cases, additional conductive hearing loss was noted.

50.8 Outcome

SK is satisfied with the FIHD on her right ear. She uses the FIHD for approximately 10 hours per day. SK also uses the M75 on her left ear for better localization and speech recognition in noise. Currently, SK has been using the FIHD for more than 4 years. Stable unaided and aided performance has been observed. Long-term satisfaction with the FIHD is also noted in this case.

50.9 Key Points

1. Audiological, medical, psychological, and socioeconomic factors should all be considered when one is assessing a patient's candidacy for an FIHD.
2. SK's objective performance with use of the FIHD was similar to that with her M75 hearing aids. This finding would not warrant fitting the FIHD.
3. Self-reported aided benefit and satisfaction were greater with the FHID in this case, thus suggesting that self-report measures and client preferences (i.e., validation) should be measured and considered as well.
4. Clinicians should investigate the available evidence and merge this evidence with SK's preferences and goals when recommending the use of newly available technology.

Suggested Reading

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51 Importance of Sufficient Output Saturation Sound Pressure Level at 90 dB on Hearing Aids

Francis Kuk

This patient reported a lack of loudness, muffled sound quality, and poor speech recognition in noise.

51.1 Clinical History and Description

JR is a 25-year-old male with a flat 60 dB HL sensorineural hearing loss. He has been wearing behind-the-ear (BTE) hearing aids with skeleton, nonvented earmolds since 3 years of age. His first hearing aids were bilateral Widex ES8 BTEs (Widex USA, Inc., Hauppauge, NY). He was fitted with bilateral Diva 9 BTE hearing aids (Widex) approximately 10 years ago. JR reported satisfaction with the Diva hearing aids, but because he was starting a new job as an engineer he thought it was prudent to upgrade the Diva 9 aids to more current technology.

After a conventional audiological examination confirmed the magnitude of hearing loss and medical clearance was obtained, the audiologist recommended bilateral digital wireless Widex CLEAR 440–9 BTEs for JR to be consistent with what he had been wearing. JR felt, however, that cosmetics were important and decided instead to purchase bilateral Widex CLEAR 440 C4-Passion (C4-Passion) receiver-in-the-canal (RIC) hearing aids. This is a subminiature RIC-style hearing aid with a thin wire connecting the hearing aid case to the receiver, which is placed inside an instant-fit dome. No vent was used on the dome.

The fitting of the C4-Passion hearing aids was unremarkable. After fitting, the audiologist verified that the simulated frequency output of the C4-Passion matched the simulated output of the Diva 9 at input levels of 50 dB and 65 dB SPL. The features on the hearing aids included Speech Enhancer Noise Reduction, active feedback cancellation, and the fully multichannel adaptive directional microphone.

JR reported satisfaction with the C4-Passion hearing aids, stating that the audiologist's speech was comfortably loud and clear. However, during the 1-week follow-up, JR reported that speech received with the C4-Passion was too soft in most situations other than very quiet listening situations. In addition, JR reported that recognizing speech in a noisy background required greater effort than before. In many situations, JR stated he was unable to recognize speech because "speech and noise all mingled together."

51.2 Questions to the Reader

1. Could a wireless technology aid contribute to these complaints?
2. Is it possible that a wearer reports satisfaction of hearing aids fitted in the clinic but not in daily listening environments?
3. Based on the patient's history and results of the fitting, would an audiologist expect dissatisfaction with the initial fit?
4. What aspects of a hearing aid could contribute to the complaint of "not loud enough" and "poor speech in noise"?

51.3 Discussion of Questions to the Reader

1. Could wireless technology contribute to these complaints?
   Wireless connectivity in hearing aids allows the hearing aids of a bilateral fit to communicate with each other. This provides the potential for automatic "up-to-the minute coordination" and adjustment of the hearing aid parameters to match the changing acoustic conditions. Thus the output of the hearing aids is the result of not only the acoustic input to the hearing aids but also the wireless input from the opposite ear. If the wireless input is not received, the gain parameters (and other settings) on the hearing aids would remain at the previous settings until the next wireless communication between the two hearing aids occurs. No loss of operation of the hearing aids (such as the hearing aids turning off or reducing gain) would occur. Thus wireless failure, even when it occurs, typically does not result in reduced loudness and poorer speech recognition in noise.
2. Is it possible that a wearer reports satisfaction of hearing aids fitted in the clinic but not in daily listening environments?
   The clinic environment is very different from the patient's daily listening environments. Thus the input to the patient's hearing aids could also be very different between the two listening environments. In other words, the scope of the fitting during a structured clinic environment is limited and may not be fully reflective of the real-world performance. Although many investigators have attempted to bring the patient's real-world listening environments into the clinic for verification, such a goal is not feasible because of the vast number of diverse listening environments and the cost to replicate such settings. Such limitations mandate that audiologists validate fittings (such as immediate follow- up) to determine if the hearing aids are beneficial to the patient.
3. Based on the patient's history and results of the fitting, would an audiologist expect dissatisfaction with the initial fit?
   At the end of the fitting, the simulated output from the Diva 9 aids was identical to the C4-Passion hearing aids. Because the patient had been satisfied with the Diva 9 aids, and there were more advanced features on the C4-Passion than on the Diva 9, one would expect JR to be equally satisfied or to express greater satisfaction with the C4-Passion.

   JR, however, had been wearing moderate-gain BTE hearing aids for over 20 years. A RIC-style hearing aid has its own acoustic characteristics, which are different from a conventionally coupled BTE. This sudden change to a different set of acoustic characteristics could impact the patient's perception of loudness and speech clarity. From that perspective, it is possible that JR may be dissatisfied with the C4-Passion. These acoustic differences include the following:

- BTEs, because of the tubing, earhook, and damper that are used, have their own resonance characteristics, which are not present in RIC hearing aids. RIC devices do not have tube resonance, other than the small tubing used at the end of the receiver. Considering that JR had been wearing conventionally coupled BTEs for over 20 years, he may have been accustomed to the resonance-enriched frequency/output response of his former Diva 9 aids.
- The Diva 9 BTE has a peak output saturated sound pressure level at 90 dB (OSPL90) of 125 dB SPL. The C4-Passion has a peak OSPL90 of 108 dB SPL (and typically below 103 dB SPL). This suggests that an output greater than 103 dB will be limited in the C4-Passion, but not in the Diva 9. Based on this difference alone, one may expect some difficulty from the patient when using the C4-Passion in some real-world environments.

4. What aspects of a hearing aid could contribute to the complaint of "not loud enough" and "poor speech in noise"?

There are principally three key functional blocks of a hearing aid that affect performance: the input stage, which includes the microphone and analog to digital converter (ADC), the processing stage, which includes the signal processing algorithms, and the output stage, which includes the OSPL90 and how output is limited.

- Input stage
    - Microphone: a microphone transduces the input from an analog signal into an electrical signal for subsequent processing. Input levels that are higher than the saturation level of the microphone (typically below 115 dB SPL) will saturate the microphone. Below the saturation level the microphone sensitivity could also affect the input going into the hearing aid. An omnidirectional microphone is equally sensitive to sounds from all directions, whereas a directional microphone is less sensitive to sounds from the sides and back. This suggests that the same input level would be less loud when it is transduced by a directional microphone rather than an omnidirectional microphone because of the lower input level. This is probably not the reason why "sounds are not quite as loud" in this case because the patient continued using a directional microphone.
    - Analog to digital converter (ADC): In a digital hearing aid, the ADC converts the electrical signal into a digital representation. In general, the more bits there are within the ADC, the greater the dynamic range. Typically, each bit can handle approximately 6 dB of the dynamic range. Using a typical bit size of 16, the typical digital hearing aid has a dynamic range of approximately 96 dB SPL. Thus, if the hearing aid converts soft sounds approaching 0 dB SPL, then the upper limit of sound that it can input will only be 96 dB SPL. Any input level more intense than 96 dB SPL would saturate the ADC and cause distortion. If the Diva hearing aid has a greater bit size or a larger dynamic range than the C4-Passion, the wearer of the C4-Passion could report that speech is not quite as loud and speech in noise is not quite as clear. In this case, both the Diva 9 and the C4-Passion use a 16-bit ADC, and both have an input dynamic range of approximately 103 to 105 dB SPL. Thus it is not likely that the patient's complaint originate from the input stage processing.
- Processing stage: Although there are many signal processing features within a hearing aid that may result in a change of loudness perception and/or sound quality, the author will restrict the discussion only to those that are common between the Diva 9 and the C4-Passion hearing aids. More specifically, these features include compression, noise reduction, and feedback management algorithms.
    - Compression: With the complaint that conversational-level sounds are not sufficiently loud, the gain parameter for average or loud input levels could be adjusted to increase its loudness. Upon examination, the gain settings insertion gain for soft sounds (IG soft) and for normal conversational level sounds (IG normal) on the C4-Passion were similar to those on the Diva 9. On the other hand, the gain settings for loud sounds in the C4-Passion were lower than the Diva 9. This may explain why sounds may not be loud enough, especially for sounds louder than a conversational level.
    - Noise reduction (NR): An NR circuit reduces the gain of the hearing aid once the input level and spectrum of the noise are determined. Different manufacturers implement NR strategies differently. In general, as the noise level increases and as the signal to noise ratio (SNR) becomes more unfavorable, greater gain reduction occurs. The typical amount of gain reduction is approximately 12 dB in Widex hearing aids, including the Diva 9 and the C4-Passion. This will result in a loss of loudness in noisy situations but will make the listening more comfortable. Because the Diva 9 and C4-Passion hearing aids use similar NR strategy, it is not likely that this algorithm in the C4-Passion is the reason for the complaints of "not loud enough" and "speech in noise not acceptable."
    - Feedback cancellation: A feedback cancellation algorithm estimates the feedback path within a hearing aid and generates a signal of the opposite phase as the feedback signal to cancel the signal before it becomes audible. In a sense, a feedback system ensures that gain is available for use and does not reduce the output. It is not likely the reason for the wearer's complaints.
- Output stage: The OSPL90 as well as the technology that is used to limit the output could affect the loudness and fidelity of the output.
    - Output limit: the OSPL90 level is limited by the receiver of the hearing aid, with a larger receiver having a higher OSPL90. When the output level exceeds the OSPL90 setting, the output will not increase regardless of the input level or gain setting on the hearing aid. The setting of the OSPL90 is typically adjusted two ways. First is the use of a default setting that is based on the average relationship between the loudness discomfort level (LDL) and the magnitude of hearing loss. Many prescriptive formulae such as National Acoustics Laboratories (NAL) and desired sensation level (DSL) have such recommendations. Also, the relationship reported by Pascoe has been used by many manufacturers as the default. Another approach is for clinicians to measure the LDL of the patient and then adjust the OSPL90 below the LDL of the patient. In principle, setting the OSPL90 below the measured individual LDL has significant theoretical merit. In practice, variability (and lack of reliability) of the LDL measures could result in an unreliable LDL measure.
        - An OSPL90 setting that is higher than the wearer's LDL could lead to loudness discomfort at moderate input

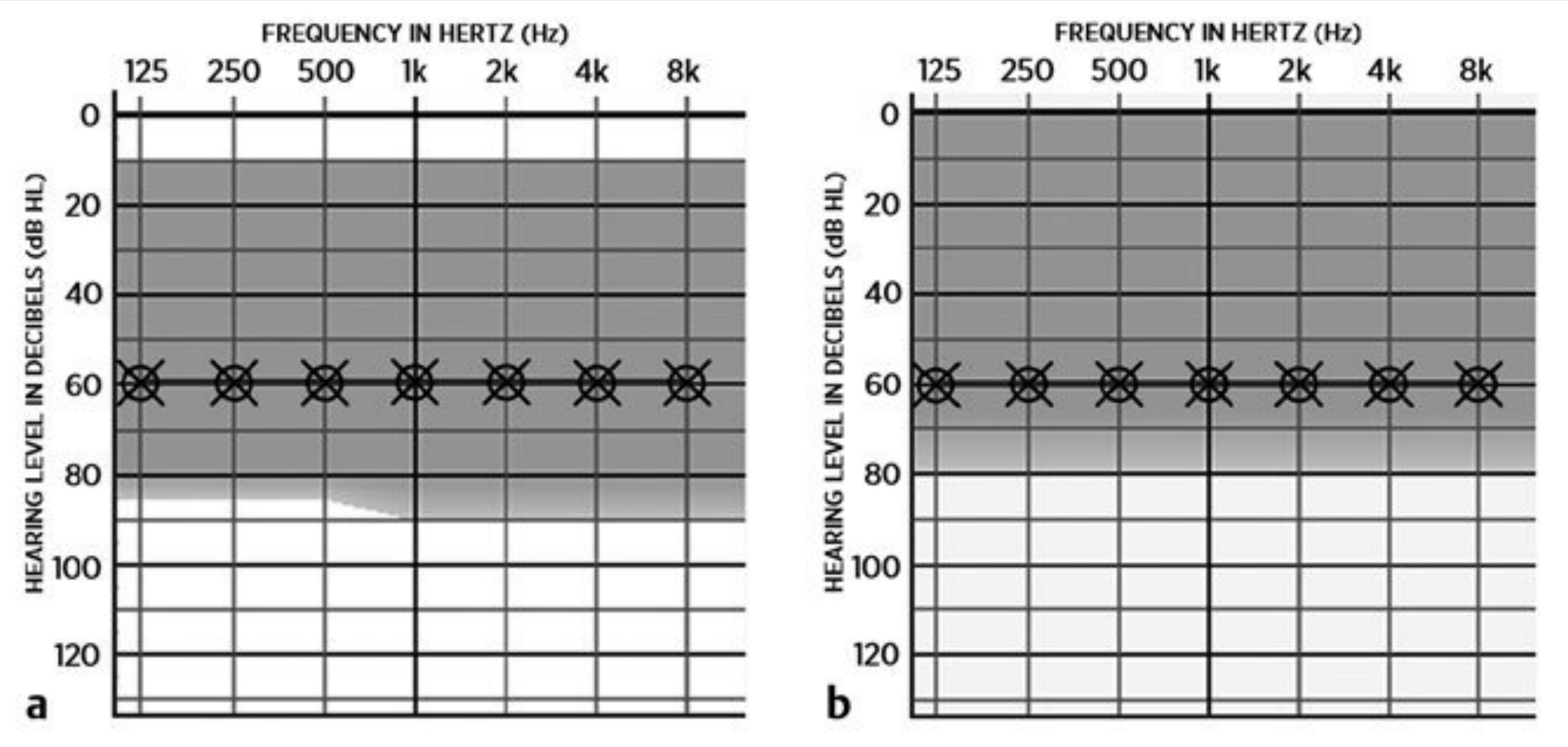

Fig. 51.1 Fitting range of the Diva 9 (Widex) (a) and the C4-Passion (Widex) (b) hearing aids.

levels. This would not only affect the use of the hearing aid but could also result in threshold shifts in hearing. Thus many audiologists take a conservative approach in setting the OSPL90 in order to minimize the risk of dissatisfaction and/or additional hearing loss. On the other hand, setting an OSPL90 that is too low (by 10 dB or more) could limit the needed output for the wearer and reduce the output dynamics of the output signal (i.e., temporal smearing). This could affect the loudness of sounds, especially those at or above a conversational level. Because of the reduced dynamics, speech and the noise background may become less differentiated to yield a muffled sound quality.

- Kuk et al demonstrated the importance of OSPL90 on speech recognition in noise. Eleven adults with severe sensorineural hearing loss participated. A single-blinded, factorial repeated-measures design was used to study the effect of noise input level (68 dB-C; 75 dB-C), OSPL90 setting (default; default-10), and noise reduction algorithm (off; classic; speech enhancer [SE]) on hearing in noise test (HINT) performance. Participants were fit bilaterally with Widex Mind 440–19 BTE hearing aids in the default frequency response and OSPL90 settings. The hearing aids were adjusted to six OSPL90 (default, default-10) by noise reduction (off, classic, SE) conditions. The reception threshold for sentences (RTS in dB) for 50%correct on the HINT was measured in each of the six conditions at two input levels of 68 dB-C and 75 dB-C with speech and noise presented from the front. A repeated-measures analysis of variance (ANOVA) revealed that noise level was not significant, whereas the noise reduction algorithm and OSPL90 were significant. The interaction between noise level and NR algorithm was also significant. The study concluded that an OSPL90 that was 10 dB lower than the default could negatively affect the SNR of the listening environment.
- There are two possibilities where the OSPL90 can be adjusted improperly. One is the measurement errors that may occur during individual measurements. The other is fitting a patient with a hearing loss that is at the extreme or beyond the fitting range of the hearing aid. This impact is more noticeable for experienced wearers. This is a likely and frequent occurrence because many wearers with more than a moderate degree of hearing loss also prefer the cosmetic appeal of smaller hearing aids such as a completely in-the-canal (CIC) or an RIC such as the C4-Passion. Although JR's hearing loss is within the fitting range of the Diva 9 (Fig. 51.1a) and the C4-Passion (Fig. 51.1b), the reader can see that JR's hearing loss is more toward the limit of the C4-Passion fitting range than the Diva 9. Although sufficient gain can be provided from both hearing aids, the available OSPL90 on the C4-Passion is lower than that on the Diva 9. This could have contributed to the "muffled speech" or "speech not loud enough" complaints.
- Output-limiting options: When the output of the hearing aid exceeds its OSPL90, it is limited to using one of two output-limiting methods. Peak clipping removes the output of the hearing aid exceeding the OPSL90. This provides an instantaneous control of the output to ensure comfort. Peak clipping, however, creates significant saturation distortion, which could affect sound quality and speech recognition. Another method is compression limiting. This reduces the gain/output of the hearing aid using a high compression ratio (>10:1). This helps limit the output and minimizes any spectral distortion. Whereas earlier forms of compression limiting were broadband in nature (all frequencies are affected simultaneously), recent advances in multichannel technology allow the use of narrow-band output limiting. This allows the hearing aid to limit the output only in the specific frequency bands that exceed the OSPL90, sparing the other frequencies from gain reduction. This could preserve the output or loudness of the hearing aid in situations where there is frequency-specific noise. The Diva 9 only has a broadband compression limiter, but the C4-Passion has both a broadband and a narrow-band compression limiter. It is likely that even in situations where a narrow-band noise is present that exceeds the output limit, the output of the C4-Passion should be higher (instead of lower) than or similar to that of the Diva 9 (if both have the same OSPL90 level). Thus the difference in the method of output limiting between the two hearing aids could not be the reason for the complaint.

51.4 Diagnosis and Recommended Treatment

Based on the previous discussions, it appears that the most likely reason for JR's complaints originated from the lower

OSPL90 and the lower default gain setting for loud sounds on the C4-Passion. Unfortunately, neither the OSPL90 setting nor the gain settings for loud sounds can be increased further because of the limit of the hearing aid. It would be optimal if JR agreed with the C4–9 BTE because it has the same electroacoustic characteristics as the Diva 9, a hearing aid JR accepted. JR's cosmetic concerns, however, need to be considered. The Clear Fusion (C4-FS) (Widex) hearing aids with the P-receiver were recommended to JR as an alternative. This is a miniature RIC hearing aid that uses a larger receiver (and thus a higher OSPL90) than the C4-Passion hearing aid. It has an OSPL90 between 118 and 124 dB SPL, which is comparable to the Diva 9. In addition, it allows additional gain adjustment for loud sounds.

51.4.1 Additional Testing

To determine if the new C4-FS hearing aids improved sound quality and speech recognition in noise, the patient was evaluated with the Diva 9, C4-Passion, and C4-FS hearing aids. Speech in noise was measured with the HINT with the sentences presented from 0° at 1 meter. An overall noise level of 70 dB SPL was presented from 90°, 180°, and 270° at 1 meter. The three hearing aids were tested in the directional mode. The HINT sentences varied in 2 dB steps after the fourth sentence. The results revealed that the Diva 9 required an RTS of − 2 dB for 50% performance. The C4-Passion required an RTS of 2 dB, and the C4-FS required an RTS of − 3 dB for 50% performance. It is important to note that the lower (or more negative) the RTS, the better the performance in noise. Assuming that each dB improvement corresponds to 8 to 10% sentence recognition, the 4 to 5 dB SNR change could result in 30 to 50% improvement in sentence recognition.

51.5 Outcome

JR reported an improvement in sound quality. Speech with the C4-FS was reportedly clearer and more natural than the Diva 9 and C4-Passion. The patient also reported a more three-dimensional perception of his listening environments. JR was seen the next day and again in 2 weeks. He confirmed his initial satisfaction with the Fusion and reported excellent sound quality and better speech recognition in noisy background.

51.6 Key Points

1. Clinicians should strive to obtain a better understanding of the wearer's previous hearing aid history and its impact on the wearer's accepting or rejecting a new hearing aid.
2. Clinicians should recognize that OSPL90 affects sound quality and speech in noise ability.

Suggested Reading

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52 An Unconventional Monaural Hearing Aid Fitting

Rachael Mangiore and Kristi Oeding

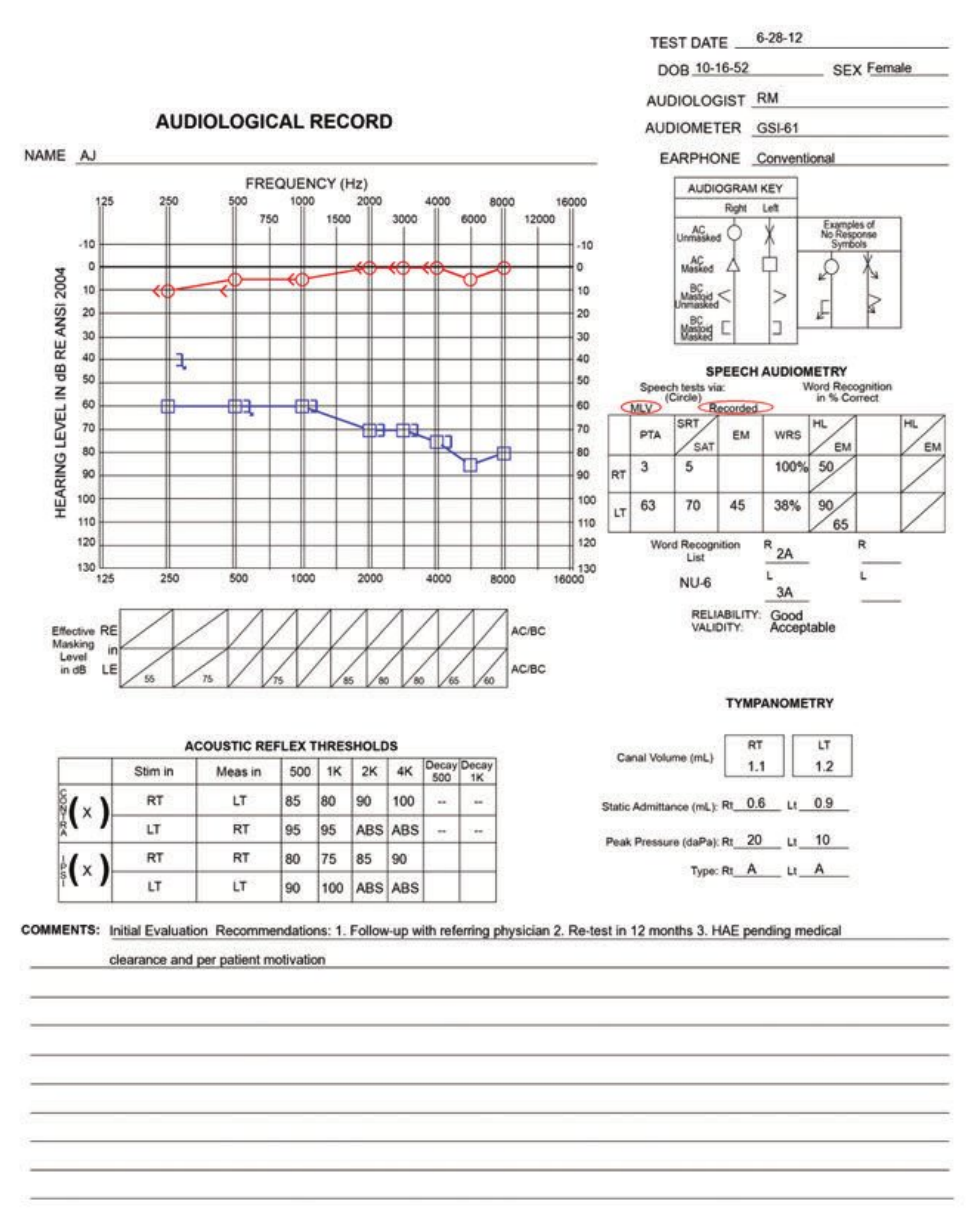
This case report summarizes the care provided to a patient with unilateral, sudden, moderately-severe to severe sensorineural hearing loss (SNHL) and very poor word recognition scores (WRSs) in the left ear and normal hearing in the right ear. Due to the patient's reported difficulty localizing sound, recognizing speech in noise, and recognizing speech on her left side, the patient inquired about amplification.

52.1 Clinical History and Description

The patient was initially seen at our clinic for a visit following a sudden hearing loss in the left ear that occurred approximately 8 years ago. The patient reported the hearing in the left ear decreased significantly in combination with an episode of vertigo requiring hospitalization. The vertigo resolved two days after use of a medication that the patient could not recall. At the time, a magnetic resonance imaging (MRI) scan was performed and was negative for retrocochlear pathology. Following the MRI scan, the patient's physician informed the patient that there was no further medical intervention to improve the hearing in the left ear. There was no follow-up medical care until the patient scheduled an appointment at our clinic. At the appointment, the patient denied recurrent vertigo since the initial episode, and there was no additional improvement of hearing in the left ear. In addition to the previous history, the patient reported constant tinnitus in the left ear that began approximately 1 year ago. The patient stated she is a very active runner and has a very difficult time localizing sound. The patient has special concerns regarding the triathlons in which she participates because she is unable to orient herself to her surroundings.

52.2 Audiological Testing

The patient completed a comprehensive audiological examination (◻ Fig. 52.1). Pure-tone thresholds revealed normal hearing



TEST DATE 6-28-12
DOB 10-16-52 SEX Female
AUDIOLOGIST RM
AUDIOMETER GSI-61
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME AJ

SPEECH AUDIOMETRY

Speech tests via: (Circle) [✓ MLV] [✓ Recorded]
Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	3	5		100%	50		
LT	63	70	45	38%	90 / 65		

Word Recognition List NU-6
R 2A
L 3A
R
L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.1	1.2

Static Admittance (mL): Rt 0.6 Lt 0.9
Peak Pressure (daPa): Rt 20 Lt 10
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	85	80	90	100	--	--
	LT	RT	95	95	ABS	ABS	--	--
IPSI (X)	RT	RT	80	75	85	90		
	LT	LT	90	100	ABS	ABS		

COMMENTS: Initial Evaluation Recommendations: 1. Follow-up with referring physician 2. Re-test in 12 months 3. HAE pending medical clearance and per patient motivation

Fig. 52.1 Initial comprehensive audiological examination.

from 250 to 8000 Hz in the right ear and a gradually sloping moderately-severe to severe SNHL from 250 to 8000 Hz in the left ear. Speech recognition thresholds (SRTs), obtained using monitored live voice (MLV), indicated normal ability to receive speech in the right ear and a moderately-severe loss in the ability to receive speech in the left ear. WRSs were obtained using recorded Northwestern University Auditory Test Number 6 (NU-6) word lists with a female talker. WRSs indicated normal ability to recognize speech in the right ear and very poor ability to recognize speech in the left ear.

Tympanometry revealed normal middle ear pressure, static compliance, and ear canal volume bilaterally. Acoustic reflex thresholds (ART) were within normal limits for right ipsilateral and contralateral stimulation at 500, 1000, 2000, and 4000 Hz and were within normal limits for left ipsilateral and contralateral stimulation at 500 and 1000 Hz. ARTs were absent for left ipsilateral and contralateral stimulation at 2000 and 4000 Hz. Contralateral acoustic reflex decay at 500 and 1000 Hz was negative (normal) bilaterally.

52.3 Questions to the Reader

1. What options for amplification are available for a patient with severe unilateral SNHL and normal hearing in the opposite ear?
2. What factors should be considered when recommending the most appropriate amplification?
3. What are the advantages and disadvantages to monaural amplification fitted to the patient's left ear?
4. What are the advantages and disadvantages to "bypassing" the left ear with a transmitting microphone?
5. With a unilateral SNHL, what are some important points to counsel the patient on during the hearing aid evaluation (HAE)?

52.4 Discussion of Questions to the Reader

1. What options for amplification are available for a patient with severe unilateral SNHL and normal hearing in the opposite ear?
   Amplification options available for a patient with unilateral SNHL are a auditory osseointegrated implant system (AOIS), SoundBite (Sonitus Medical, San Mateo, CA), contralateral routing of the signal (CROS), TransEar (Ear Technology Corp., Johnson City, TN), transcranial CROS, and monaural amplification in the poorer ear.

   The AOIS for this type of hearing loss is a device in which a titanium screw and abutment are surgically implanted into the mastoid process on the side of the poorer ear. After a three month waiting period for osseointegration of the titanium screw to occur, a sound processor is coupled to the implanted abutment to stimulate the cochlea of the better ear via bone conduction.

   The SoundBite is also based on transmitting sound to the cochlea of the better ear via bone conduction. With a SoundBite there are two devices, a transmitting microphone embedded in a hearing aid worn over the poorer ear and a frequency modulated (FM) receiver molded into a retainer sitting adjacent to the molars of the patient on the side of the better ear. The FM transmitting microphone wirelessly transmits the signal from the poorer ear to the FM receiver placed in the retainer in the patient's mouth next to the molars, which stimulates the cochlea of the better ear via bone conduction.

   CROS amplification places a transmitter microphone on the poorer ear and a receiving hearing aid on the better ear. The sound is wirelessly transmitted to the receiver hearing aid on the better ear by amplitude modulation (AM) or FM sound transmission. The receiver hearing aid is coupled to the ear using either a custom open earmold or a generic open dome to allow for natural sound propagation into the ear canal because these patients have normal or near normal hearing in the better ear.

   The TransEar is a bone conduction hearing aid in which a custom earmold sits deep in the ear canal of the poorer ear. A behind-the-ear (BTE) sound processor connects to the earmold via an appropriate size wire. An oscillator inside the earmold makes contact with the bony portion of the ear canal. The amplification provided by the processor to the bone conduction vibrator sitting inside the earmold must be sufficiently loud that the sound will be received by the cochlea of the better ear.

   The transcranial CROS is a conventional air conduction hearing aid in which the amplified signal is presented to the poorer ear and the amplified sound must be sufficiently loud enough to cross over via bone conduction to the cochlea of the better. The patient's hearing levels in the better ear along with his or her individual interaural attenuation and transcranial thresholds determine the output that is appropriate to reach the cochlea of the better ear.
2. What factors should be considered when recommending the most appropriate amplification?
   When recommending the most appropriate amplification to a patient, there are several factors to consider. The audiologist should account for the magnitude and configuration of the hearing loss, lifestyle of the patient, physical limitations, comfort of the patient, and the patient's needs and wants. The device must be able to provide the appropriate amount of gain/output for the hearing loss that is present. Also, all the features that are required to best benefit the patient must fit within the hearing aid. These features may include dual microphones, noise reduction, feedback management, number of channels and bands of signal processing, telecoil, and so on. Once the appropriate amount of amplification is determined and the physical requirements of the hearing aid are met, the patient's preferences must be considered. In the case of the present patient, cosmetics and size were not important considerations. The patient was interested in being fit with a device that provided her with the best benefit. Her primary goal was to achieve sound awareness and the ability to localize sound. The only option that met those two requirements was monaural amplification to the poorer left ear.
3. What are the advantages and disadvantages to monaural amplification fitted to the patient's left ear?
   The advantages of monaural amplification to the left (poorer) ear versus CROS amplification include elimination of the head shadow effect, sound localization, and possible

prevention of auditory deprivation in the event a cochlear implant (CI) becomes an option for this patient in the future. The disadvantages to monaural amplification to the left (poorer) ear versus CROS amplification include the fact that, in this case, amplifying distorted speech due to an ear with very poor word recognition (38%) could be distracting and detrimental for the patient.

4. What are the advantages and disadvantages to "bypassing" the left ear with a transmitting microphone?
   The advantages to CROS amplification to the better ear compared with monaural amplification to the left (poorer) ear include elimination of the head shadow effect and sound awareness (a clear signal as it is being sent to the normal hearing ear) on the side of the poorer ear. The disadvantages to CROS amplification versus monaural amplification to the left (poorer) ear include possible auditory deprivation and implications for future cochlear implantation. Although its significance has not yet been determined, it has been suggested that an indicator for successful cochlear implant outcomes is the length of time that amplification has been used in the hearing impaired ear in relation to the length of time the hearing loss has been present. The theory is that the more time spent using amplification prior to implantation the more successful the user's predicted outcome will be.
5. With a unilateral SNHL, what are some important points to counsel the patient on during the hearing aid evaluation (HAE)?
   When counseling a patient with unilateral SNHL and poor WRS it is important to present the advantages and disadvantages of the various fitting options during patient counseling. In addition to those factors, it is important to discuss patient expectations. As the audiologist, it is important to provide reasonable expectations with any amplification. In this case, it is imperative that the patient understand the advantages and disadvantages to each available amplification option regarding speech recognition, localization, and future benefit from amplification if her hearing loss in either ear should decline.

52.5 Diagnosis and Recommended Treatment

The patient was referred to her physician regarding the sudden hearing loss and vertigo and a consultation with an otologist was recommended to rule out a retrocochlear pathology. It was also recommended that an auditory brainstem response (ABR) test be completed as well as an HAE following medical clearance for amplification. Her physician did not order a repeat MRI scan and ABR because the physician determined that the patient had experienced SNHL in the left ear with vertigo eight years prior. Her physician determined the vertigo had since fully compensated and recovered and recommended she return to her audiologist for an HAE to determine the most appropriate amplification option available. The patient returned two weeks following the audiological examination for an HAE to discuss amplification options. At this appointment, the patient was counseled regarding the advantages and disadvantages of the AOIS, SoundBite, CROS amplification, TransEar, transcranial CROS, and monaural amplification in the poorer ear. Also, her expectations with the amplification options were obtained. The patient was not concerned about sound quality and distortion as much as she was concerned about sound awareness and sound localization to her left side. The patient expressed interest in being able to use the device during triathlons and exercise.

After the extensive counseling at the HAE, the patient elected to pursue monaural amplification to the poorer left ear. A Phonak Naida S CRT V (Phonak U.S., Warrenville, IL) with a power receiver and custom c-shell mold were chosen and ordered. This particular hearing aid was selected, among other reasons, for the moisture-resistant coating it maintains in order for the patient to use the hearing aid during triathlons and exercise. She was also provided with a Zephyr by Dry & Store (Ear Technology Corp., Johnson City, TN) for additional protection from moisture-related damage of the hearing aid. The patient understood that this is an unconventional option for a patient with very poor speech recognition in the ear to be aided. She also understood that speech recognition may be negatively affected by providing amplification to the ear with very poor speech recognition as compared to CROS amplification, which would "bypass" the ear with very poor speech recognition. The hearing aid fitting was performed two weeks following the HAE after the patient had received medical clearance for amplification from her physician. Loudness discomfort levels (LDLs in dB SPL) were obtained using real ear measures from 500 to 4000 Hz in the left ear to determine the level at which frequency-specific pure-tones cause discomfort. LDLs were obtained prior to the hearing aid fitting to verify that the real ear saturation response using a 90 dB SPL (RESR90) pure-tone sweep did not exceed the LDL at each frequency measured (Fig. 52.2b). The patient judged the loudness of the pure-tone

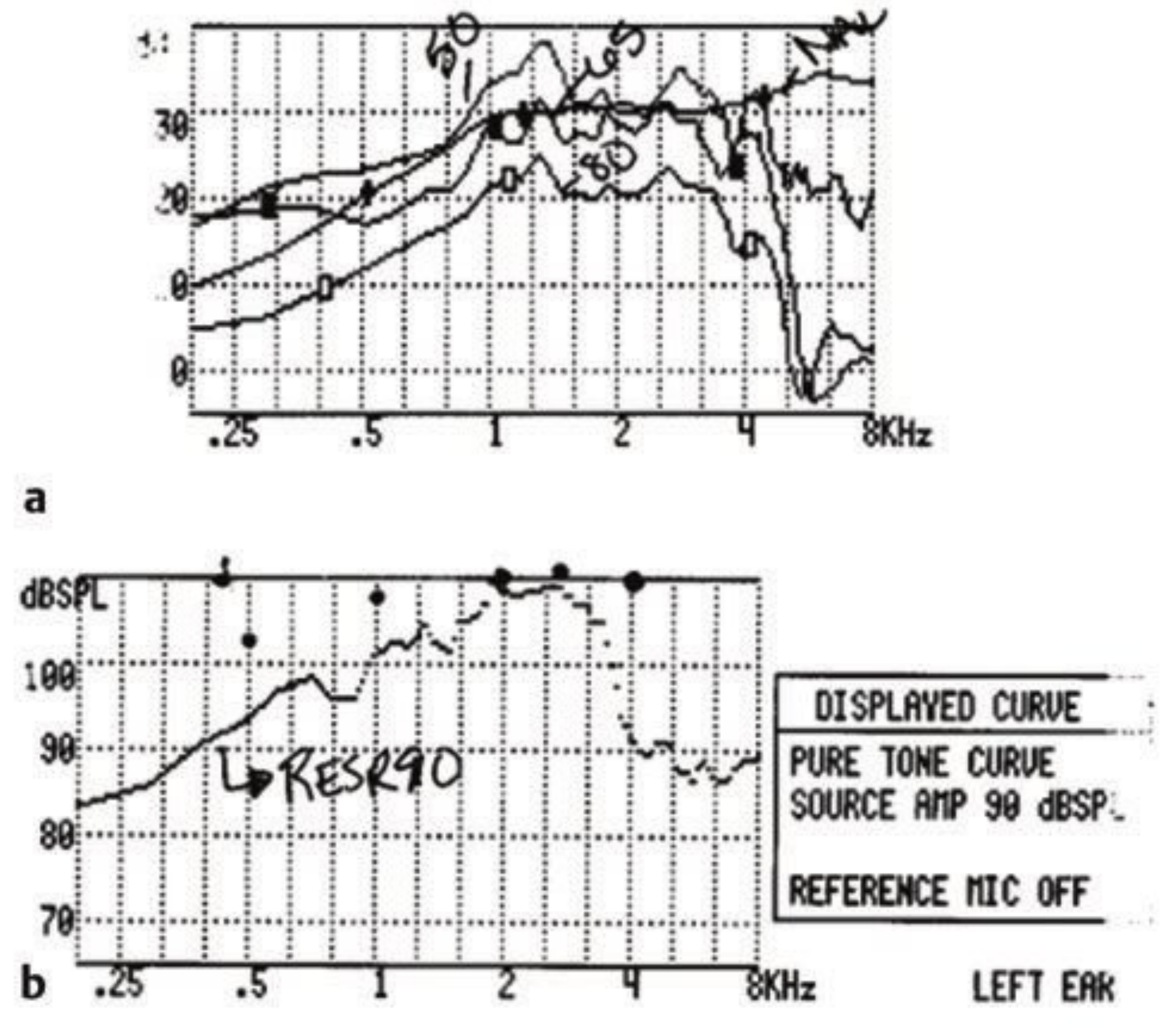


Fig. 52.2 (a) Initial hearing aid fitting using the Frye 6500 real ear analyzer (Frye Electronics, Inc., Tigard, OR) for the Naida S CRT V (Phonak) at input levels of 50, 65, and 80 dB SPL using speech-weighted composite noise as well as the real ear saturation response using a 90 dB sound pressure level (RESR90) (b). In (a), the real ear insertion gain (REIG) curves are labeled 50, 65, and 80 for each input level and NAL-NL1 prescriptive target for the 65 dB SPL input level. On (b) graph, the "dots" represent the individually measured LDLs at each measured frequency. The curve represents the RESR90.

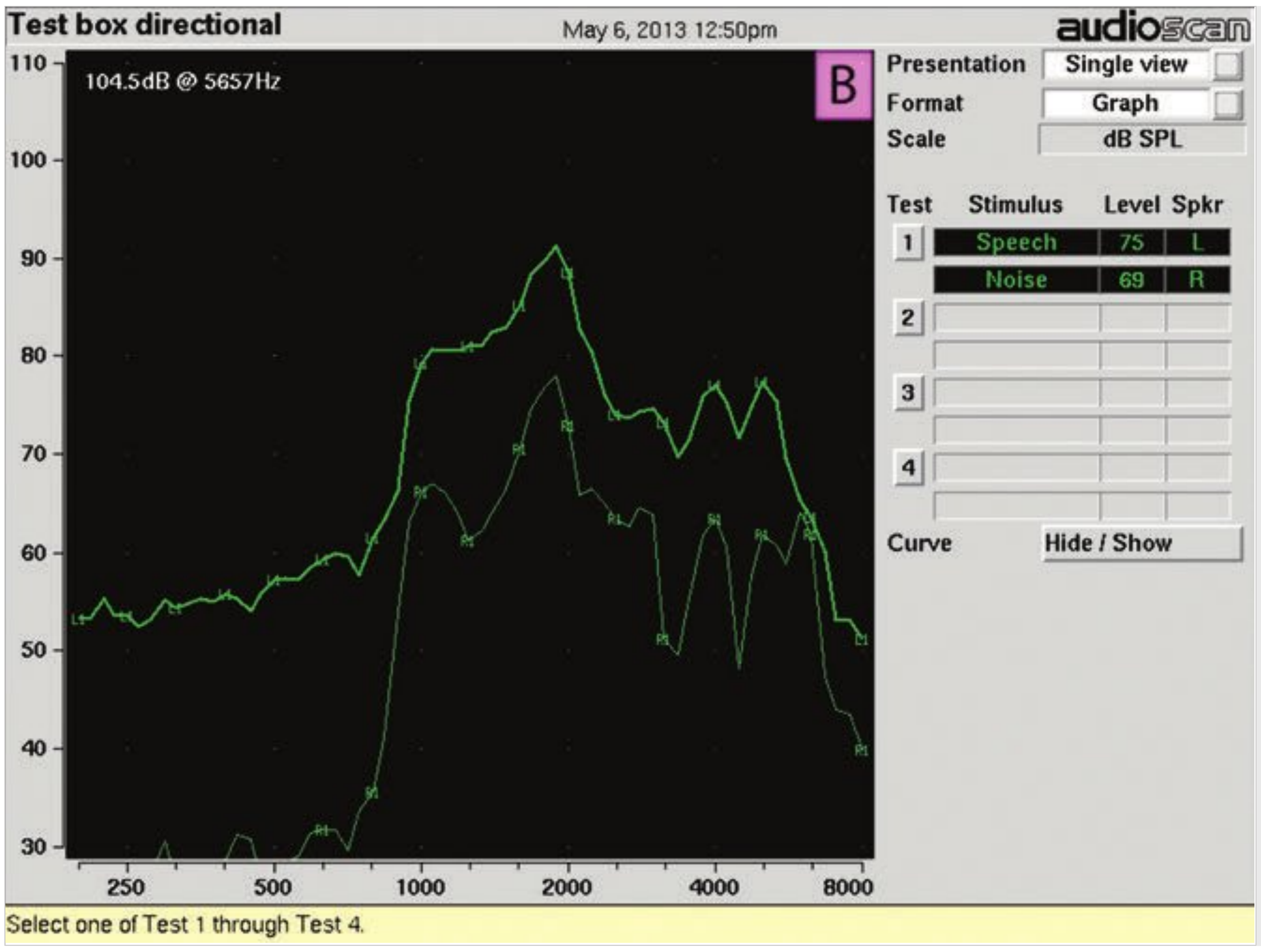
sweep as “loud, but okay” indicating that the sound was not “uncomfortably loud” or “comfortable, but slightly loud.” These loudness judgments were rated as described by Hawkins et al. In Fig. 52.2 the “dots” represent the patient’s individually measured LDL in dB SPL at each frequency and the curve represents the RESR90 from 250 to 8000 Hz. This figure illustrates that the RESR90 did not exceed the patient’s individually measured LDL at any frequency and suggests the patient should not report that the hearing aid is ever “uncomfortably loud.”

Following the measurement of the RESR90, real ear insertion gain (REIG) target values were established for the left ear using National Acoustic Laboratories’ nonlinear fitting formula version 1 (NAL-NL1) for input levels of 50, 65, and 80 dB SPL corrected for channel summation of 16 channels. Real ear measurements (REMs) for the Naida S CRT V were obtained using modulated speech-weighted composite noise (Fig. 52.2a). These measurements verified that the hearing aid provided appropriate gain at the three input levels and a smooth frequency response. Loudness judgments were also obtained for input levels of 50, 65, and 80 dB SPL using speech-weighted composite noise. The patient reported loudness judgments of “soft” for the 50 dB SPL input, “comfortable” for the 65 dB SPL input, and “loud, but okay” for the 80 dB SPL input. The performance of the directional microphones was verified using the Verifit test box (AudioScan, Dorchester, ON, Canada) dual noise signal with a 6 dB SPL signal-to-noise-ratio (SNR) comparing the amplified frequency response at 0 degrees and 180 degrees (Fig. 52.3). In Fig. 52.3, the top curve represents the frequency response from the front microphone at 0° azimuth and the bottom curve represents the frequency response from the back microphone at 180° azimuth. Frequency from 250 to 8000 Hz is located on the abscissa and decibel level in SPL is located on the ordinate. The top curve reports that the front microphone is providing more output than the lower curve across all frequencies in the presence of noise. The difference between these two curves is known as the front-to-back ratio and reports how effective the directional microphone is performing to reduce amplification from behind. This measure is particularly important to perform due to the possibility of receiving a purchased hearing aid from the manufacturer with the directional microphones either reversed or not functioning at all. The patient was then counseled extensively on the insertion and removal of the hearing aid, use and care of the hearing aid, loss, damage and repair warranties, battery use and replacement, and the trial period with the hearing aid. The patient understood that she had time to use the hearing aid during the trial period in her natural listening environments prior to deciding to keep the hearing aid, return the hearing aid, or exchange it for another amplification option.

Follow-up to the hearing aid fitting included a phone call one week after the fitting and a hearing aid assessment (HAA) approximately two weeks following the hearing aid fitting. At the HAA, the patient is asked to describe any observations made of comfort, sound quality, and function of the hearing aid including battery life and possible intermittencies, so that programming or physical fit adjustments may be made to maximize the benefit of the trial period. At the one week phone call, the patient stated she was doing very well with the hearing aid and she had no complaints. At the HAA, the patient reported she was subjectively performing very well with the left hearing aid. The patient had used the hearing aid in multiple listening environments, including exercising in background noise, watching television, and in quiet. The patient stated she achieved her goal in sound awareness and sound localization to the left side and was very pleased with the hearing aid fit to her left ear. The patient also noted that while she was wearing the device on the left ear, the tinnitus was no longer present, which allowed the patient to better concentrate in quiet environments.

52.6 Outcome

To date, the patient remains satisfied with the left hearing aid. The patient reported improved sound awareness to her left


Test box directional

May 6, 2013 12:50pm

104.5dB @ 5657Hz

Presentation: Single view

Format: Graph

Scale: dB SPL

Test	Stimulus	Level	Spkr
1	Speech	75	L
	Noise	69	R
2			
3			
4			

Select one of Test 1 through Test 4.

Fig. 52.3 The front-to-back ratio of the Naida S CRT V (Phonak) using the Verifit® hearing aid test box (AudioScan) to verify the performance of the directional microphones.

side, improved ability to localize sound in her environment, and reduced awareness of the tinnitus while wearing the hearing aid. The patient is scheduled for follow-up hearing aid maintenance as well as audiological examinations to monitor hearing bilaterally.

52.7 Key Points

1. Counseling on user expectations from amplification is important. Each patient is different, and it is essential that he or she be aware of the advantages and disadvantages of each amplification option that is presented in order to properly and appropriately determine the patient's expectations.
2. The typical patient would not be expected to perform as well with a hearing aid amplifying an ear with poor word recognition, which leads to distortion in the poorer ear as compared with CROS amplification. As demonstrated in this case, the needs and expectations of the patient are key components in choosing the most appropriate amplification.

Suggested Reading

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53 Hearing Aid Candidacy and Selection

Lisa Lucks Mendel

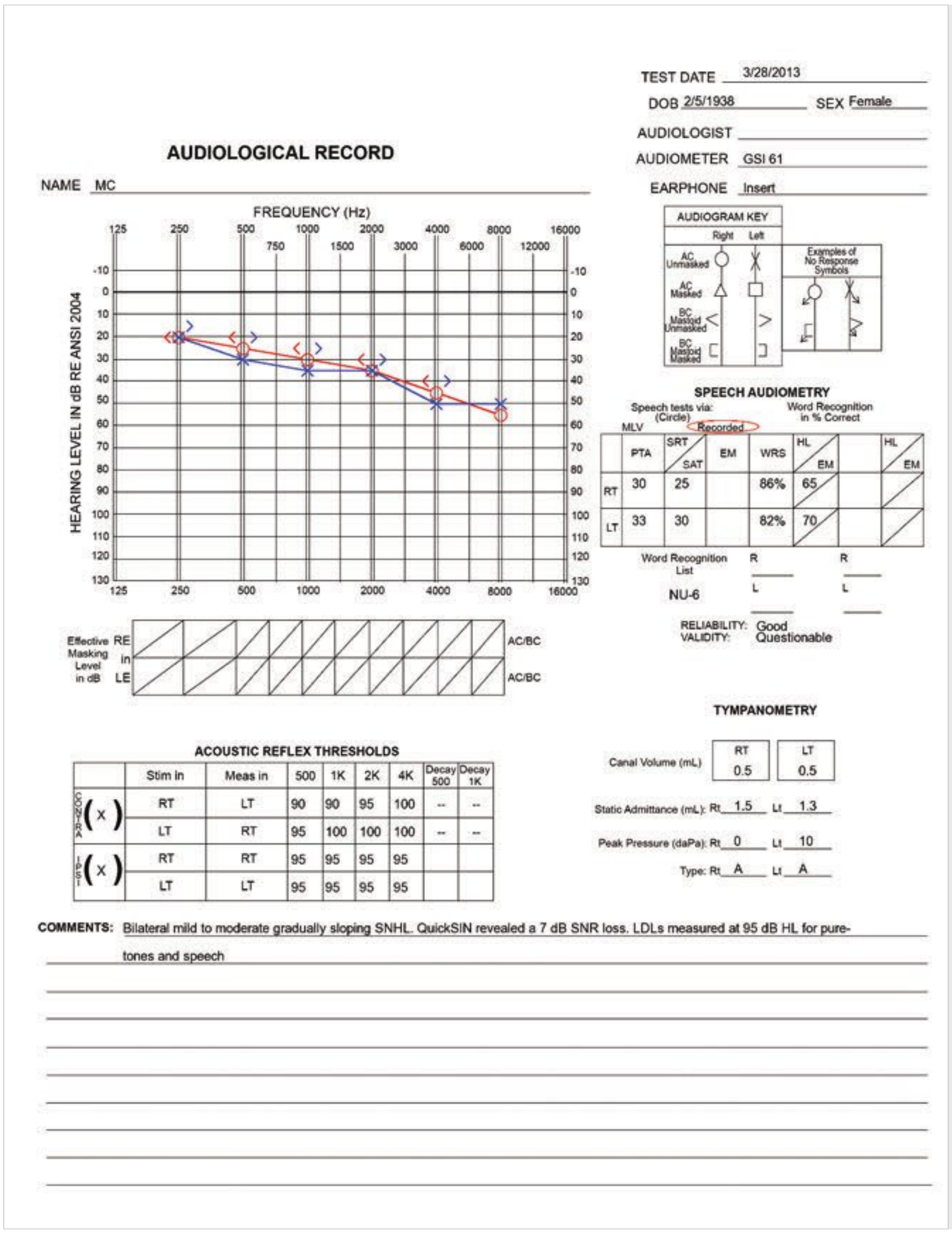
This case report addresses some of the major issues that must be considered in determining hearing aid candidacy as well as acoustic and nonacoustic factors to consider in hearing aid selection.

53.1 Clinical History and Description

MC is a 75-year-old female who scheduled an appointment in the audiology clinic. MC is accompanied by her son who reports his mother's hearing has deteriorated to the point that he is having greater difficulty communicating with her, and he is concerned about her living alone. When asked, MC admits she sometimes has trouble hearing her grandchildren and occasionally struggles hearing her friends at her bridge club. She enjoys watching television, going to the symphony, and gardening. She also reports she has arthritis in her fingers that is bothersome when the weather is bad.

53.2 Audiological Testing

Audiometric examination revealed a bilateral mild to moderate gradually sloping sensorineural hearing loss (Fig. 53.1). Speech recognition thresholds (SRTs) were 25 and 30 dB HL in the right and left ears, respectively, indicating a mild loss in the ability to receive speech; SRTs were in good agreement with the pure-tone averages. Word recognition scores (WRSs) were obtained using 50-item Northwestern University Auditory Test No. 6 (NU-6) word lists recorded by a male talker presented at 40 dB sensation level (SL) re: SRT. WRSs reflected a slight loss in the ability to recognize speech in quiet bilaterally (86% right; 82% left). Administration of the QuickSIN (Etymotic Research, Inc., Elk Grove Village, IL) revealed a 7 dB signal-to-noise ratio



AUDIOLOGICAL RECORD

NAME MC

TEST DATE 3/28/2013

DOB 2/5/1938 SEX Female

AUDIOLOGIST

AUDIOMETER GSI 61

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT/SAT	EM	WRS	HL/EM		HL/EM
RT	30	25		86%	65		
LT	33	30		82%	70		

Word Recognition List NU-6 R L R L

RELIABILITY: Good
VALIDITY: Questionable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.5	0.5

Static Admittance (mL): Rt 1.5 Lt 1.3

Peak Pressure (daPa): Rt 0 Lt 10

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	90	90	95	100	--	--
	LT	RT	95	100	100	100	--	--
IPSI (x)	RT	RT	95	95	95	95		
	LT	LT	95	95	95	95		

COMMENTS: Bilateral mild to moderate gradually sloping SNHL. QuickSIN revealed a 7 dB SNR loss. LDLs measured at 95 dB HL for pure-tones and speech

Fig. 53.1 Audiometric findings.

(SNR) loss bilaterally indicating that MC needs a listening environment with a +7 dB SNR to perform as well as a listener with normal hearing. Loudness discomfort levels (LDLs) were measured at 95 dB HL bilaterally to pure-tones presented at 500, 1000, 2000, and 4000 Hz and speech. Immittance audiometry revealed Jerger type A tympanograms bilaterally with normal ear canal volume, tympanometric peak pressure, and acoustic admittance suggesting normal middle ear function. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present at reduced sensation levels bilaterally, consistent with the audiometric findings; acoustic reflex decay was negative bilaterally.

53.3 Questions to the Reader

1. What factors need to be addressed to determine if MC is an appropriate candidate for hearing aids? How will this information be determined?
2. What are some potential obstacles that MC might experience in acquiring hearing aids?
3. What considerations need to be addressed regarding the electroacoustic characteristics, compression parameters, and special features to be included in the hearing aid(s)? How will this information be determined?
4. What nonacoustic features should be considered in the selection of hearing aids for MC? Could MC benefit from hearing assistive technology (HAT)?

53.4 Discussion of Questions to the Reader

1. What factors need to be addressed to determine if MC is an appropriate candidate for hearing aids? How will this information be determined?
   MC's initial motivation and level of acceptance of her hearing loss will impact her pursuit of and continued use of amplification. Therefore, the clinician must first explore motivational issues and communication needs. For example, has MC acknowledged she has a hearing loss or does she blame her communication problems on the fact that "other people mumble?" The clinician must also determine what MC's communication needs are and whether she has any specific activity limitations and/or participation restrictions that are a direct result of her hearing loss. To do this, a structured interview could be most helpful at getting an in-depth profile of MC's needs. In addition, using a standardized outcome measure such as the Hearing Handicap Inventory for the Elderly (HHIE) would provide the clinician with specific information regarding MC's self-reported disability. Another useful tool is the Self-Assessment of Communication (SAC), which addresses primary communication difficulties as well as secondary emotional and social consequences from the hearing loss. In addition, the SAC has a version for significant others to complete (Significant Other Assessment of Communication [SOAC]) that can provide valuable additional information for the clinician to consider regarding MC's family's perceptions of the impact of her hearing loss.

   It is also important to determine what MC's expectations are concerning amplification. Many patients have preconceived notions about amplification based on what they have been told by friends or family about hearing aids. The clinician should explore whether MC has any fear or uncertainty regarding expected benefit, costs, cosmetic concerns, or the impact the hearing aids may have on her self-image. Another useful self-report measure is the Client-Oriented Scale of Improvement (COSI), which can be used in the initial stages of the hearing aid fitting to help MC identify as many as five listening situations in which she would like to hear better. The advantage of the COSI compared with some other self-report measures is that the situations are specifically determined by the patient. The COSI can then be readministered several weeks after the hearing aid fitting to help determine if benefit in these specific areas has been achieved.

   Assessment of MC's motivation for amplification and communication needs is a fundamental, critical step in the hearing aid selection process. Keep in mind, however, that no one factor will likely suggest that a patient will or will not benefit from amplification. Therefore, it is important to counsel MC throughout this process and help her develop realistic goals and expectations as she continues to accept her hearing loss and the recommended treatment.
2. What are some potential obstacles that MC might experience in acquiring hearing aids?
   Based on the assessment information collected in this case, it is possible that MC may not be a good candidate for hearing aids. If the results from her HHIE and SAC questionnaires reflect that she has little or no self-perceived disability, it may be premature to consider amplification. She may also perceive negative stigma associated with hearing loss in older individuals. In addition, MC may have been told by her friends that hearing aids provide little help or poor quality of sound. This reinforcement of negative attitudes regarding hearing aids by friends and family may influence her not to pursue hearing aids at this time. Other potential obstacles to acquiring hearing aids at this time could include difficulty manipulating small objects, other health problems, and financial cost. If any of these complications appear to be a factor, counseling will be necessary for MC and her family.
3. What considerations need to be addressed regarding the electroacoustic characteristics, compression parameters, and special features to be included in the hearing aid(s)? How will this information be determined?
   Assuming MC has decided to pursue amplification, the clinician will need to review her audiological results, LDLs, and WRSs to make some decisions regarding how much gain will be needed, where maximum output should be adjusted, and what type of compression should be implemented. The clinician will also use information provided in the case history to make these decisions. Given that one of MC's concerns is that she has trouble hearing her grandchildren, some form of wide dynamic range compression (WDRC) should be considered because this form of compression provides more gain for soft sounds. In addition, because MC's LDLs are reasonably low, she will also need some output compression limiting (OCL) to limit output for a loud input level. Given that MC has a narrow dynamic range, the clinician should begin counseling her regarding realistic expectations about what hearing aids can and cannot provide.

   Learning more about MC's lifestyle will help determine what additional features should be included in her hearing aids. For example, MC reported she is socially active. This information assists the clinician in determining what different situational programs might be included in the hearing aids (e.g., listening in noise at her bridge club, listening at a distance for the symphony, etc.). The clinician also knows that MC is exposed to a variety of noisy listening environments, and she has trouble hearing in noise, so the clinician should consider directional microphones and possibly digital noise reduction (DNR) features in the hearing aids. The clinician may need to ask MC if she would prefer to have the hearing aids make adjustments automatically (e.g., automatic and adaptive directionality) or whether she would like to have some control (e.g., use of a remote to manually control these features with a program button and volume control). Finally, if the high-frequency slope of MC's hearing loss is significant, feedback management may be necessary.

4. What nonacoustic features should be considered in the selection of hearing aids for MC? Could MC benefit from hearing assistive technology (HAT)?
   Nonacoustic features are an essential consideration in hearing aid selection as well. MC has reported she has some arthritis, so dexterity issues could come into play. These issues may have an impact on the size and style of the hearing aids selected because a larger hearing aid using a larger battery may be easier to manipulate. In addition, the clinician may need to consider a raised volume control (if appropriate) and/or a remote control to assist MC in manipulation of the devices. Further, the clinician should discuss MC's use of the telephone to determine if a telecoil should be included in at least one of the hearing aids.

   Coupling options are an important consideration for the nonacoustic features of the hearing aids. In this case, it is likely that MC could benefit from an open fitting, so the clinician will need to consider dome options. If a custom mold is selected instead of a dome, then acoustic modifications such as venting could be of significant benefit to MC. Because MC also reports she enjoys gardening, she may need a microphone wind screen and the option to switch to an omnidirectional microphone while in noise to improve performance in windy situations. The clinician also needs to consider the use of wax guards if there is a tendency for wax to accumulate in the ear canals.

   In the case history, MC's son was concerned about his mother living alone because of her hearing loss. This concern provides an excellent opportunity for the clinician to address the possibility of HAT in addition to hearing aids. Alerting devices such as modified smoke detectors might be a consideration. An amplified phone may also be of interest to MC. In addition, perhaps some type of wireless streaming technology would provide MC with an easier ability to enjoy the television.

53.5 Diagnosis and Recommended Treatment

MC has a bilateral mild to moderate gradually sloping sensorineural hearing loss that could benefit from amplification. After discussing MC's hearing loss with her and her son, the clinician administered the HHIE, SAC, and SOAC to determine MC's communication needs, her family's perception of her needs, and her readiness for pursuing amplification. The clinician addressed many of the hearing aid selection issues already discussed here by talking with MC about the information provided in Table 53.1, which addresses the acoustic and nonacoustic issues that must be considered when selecting amplification. For each relevant question or issue, the clinician assessed MC's needs and determined an appropriate recommendation for each need. Each recommendation assisted the clinician in determining the appropriate acoustic and nonacoustic features to be ordered with the hearing aids.

53.6 Outcome

After reviewing the results of the questionnaires and the information obtained by the clinician (Table 53.1), the clinician recommended that MC be fit with bilateral mini behind-the-ear (BTE) hearing aids with open domes. The hearing aids have as many as four programs available for specific settings for various listening situations and include directional microphones, feedback management, and DNR. MC's preference was to have the hearing aids be as automated as possible so she would not need to think about the hearing aids. To provide this, automatic and adaptive directionality were included. MC did not feel she used the telephone often enough, so she decided not to have a telecoil activated in the hearing aids. The clinician counseled MC that she would still likely be able to hear well with the hearing aids over the phone using acoustic coupling. If not, she could consider purchasing a telephone amplification system.

The COSI was administered at the time the hearing aids were ordered so the clinician could document the areas where MC would like to experience improvement with the hearing aids. MC was counseled again regarding realistic expectations for the hearing aids. One of the items MC included in the COSI was the ability to hear the television better, so the clinician ordered a compatible streamer. Additional counseling will be performed while MC's son is present to ensure that MC can use the streamer appropriately in her home.

When MC returned for a 30-day follow-up, she reported she was adjusting well to her new hearing aids and finding them beneficial in most situations. Her primary complaint was difficulty hearing over the phone with her hearing aids. The clinician gave MC the opportunity to use the amplified telephone available in the clinic, and she reported it was much better than her phone at home. The clinician recommended that MC purchase an amplified telephone similar to the one available in the clinic. MC also reported that wind sounded loud through her hearing aids when she was gardening. The clinician asked if MC was using Program #4 when she gardened, and MC indicated she had forgotten about that program. The clinician determined the wind program was working well and reinstructed MC to use Program #4 when outdoors.

53.7 Key Points

1. In addition to the audiological findings, a patient's motivation for pursuing amplification and any potential obstacles must be considered when assessing hearing aid candidacy.
2. Communication needs must be assessed using interviews and/or questionnaires for the patient and his/her significant others.

Table 53.1 Considerations for choosing hearing aids for an adult; table completed by MC and the clinician.

Name: MC Patient #: 2468 Date: 3/28/2013. Please rank the following from 1 to 4 in terms of their importance to you when purchasing a hearing device. (1 = most important and 4 = least important)

1	Sound quality and clarity	1	Durability/reliability	2	Cost	2	Appearance

Question or issue	How assessed	Y/N	If yes	Recommendation
Dexterity problems?	Pill dispenser with batteries Have patient flip through a phone book. Rating: none/mod/severe	Y	Raised volume control (VC) BTE Remote control	Remote control
Previous user?	Consider case history. Ask about likes and dislikes regarding current amplification.	N	May need VC On/off switch? Counseling re: quality changes with WDRC	
Socially active?	How do you spend your day? Bridge club, symphony, and gardening	Y	Which different environments needed?	Quiet Groups Music Wind
Noisy listening environments?	Tell me about the listening situations in which you have difficulty. Are you often in situations where there is background noise?	Y	Directional microphone (automatic, adaptive?). BTE/ITC Noise management algorithm	Automatic, adaptive directional microphone
High-frequency options	Check audiogram for steeply sloping configuration or high-frequency dropoff.	N	Feedback management Frequency lowering feature	Feedback management given open fitting
Outdoor activities?	How much time do you spend outdoors? Lots What outdoor activities do you do? Gardening	Y	Noise or wind noise management Perspiration issues	Wind noise management; perspiration issues
Need/desire for automatic functioning?	Consider manual dexterity. Would you prefer to have the HA do everything for you or would you like to have some control?	Y	Willing to accept remote control? Need for VC? Push button control?	Yes—Remote Yes—VC Push button—OK
Need/desire for telephone help (landlines only)?	How often do you use the telephone? Some Do you have trouble hearing over the phone? Yes Do you use your HA with the phone? Which ear? (previous user)	Both	Discuss pros/cons of automatic telecoil. Telecoil or acoustic phone?	No autocoil Wants to try acoustic coupling
ALD or device compatibility needed?	Do you wish to use entertainment devices such as MP3, iPOD, etc.? Yes?	Do you live alone? Yes Do you use any devices to help you hear better at home or outside home? No Do you have trouble hearing the television? Yes! Do you have difficulty hearing in a large room? Yes	Alerting device(s) Telecoil or direct audio input for use with personal FM, or loop system Entertainment device compatibility, e.g., streamer	Compatible streamer
Preference for one or two? (see below)	Would you prefer one or two HAs? Two Check audiogram. Speech perception asymmetry Ear anomalies	2	Plan a strategy for allowing patient to experience monaural and binaural fittings if he/she wishes. Order appropriate hearing aids.	Wants to try two aids at first

Table 53.1 continued

Name: MC Patient #: 2468 Date: 3/28/2013. Please rank the following from 1 to 4 in terms of their importance to you when purchasing a hearing device. (1 = most important and 4 = least important)				
1 Sound quality and clarity	1 Durability/reliability	2 Cost	2 Appearance	
Research has consistently shown that even patients with bilateral symmetrical hearing loss quite often prefer to wear only one hearing aid. Preference cannot be predicted using audiological variables. Work with patient to find the preferred mode.				
Preference for HA style	Do you have an opinion about the style of the HA? Are you concerned about other people noticing it?	N	Show models and/or pictures of different styles	Open to any style
Coupling options	Check audiogram configuration, LF hearing, HF hearing, and severity of loss. Custom earmold or dome fitting?		Custom mold: Venting Horn effects Sound bore Receiver mold Dome: Open Closed	Dome tubing length Oticon__2___ Siemens__1____ Phonak__2____ Starkey__1___ Resound__2___
Wax problems?	Otoscopy Do you tend to have a lot of wax in your ears? Do you often have the wax in your ears removed?	N	Wax guard Receiver-in-the-canal (RIC) versus receiver-in-the-aid (RITA)	RIC
Ear drainage?	Otoscopy Do you have liquid draining from your ears?	N	Open fitting	
Abbreviations: ALD, Assistive Listening Device BTE, behind-the-ear; HA, hearing aid; ITC, in-the-ear. Source: Reprinted with permission. © 2011, University of Memphis.				

3. Counseling regarding realistic expectations can be beneficial for a successful outcome.
4. Information from the case history regarding primary concerns and lifestyle together with the audiological results must be considered when determining the electroacoustic characteristics needed for the hearing aids.
5. Nonacoustic features such as microphone windscreens, venting, and wax guards are important aspects of hearing aid selection.
6. HAT beyond personal amplification should be pursued with patients and families.

Suggested Reading

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54 Amplification for Mixed Hearing Loss

Kristi Oeding and Rachael Mangiore

This case report describes the challenges in fitting a patient with longstanding bilateral otosclerosis along with recent sudden hearing loss in her right ear. Several amplification treatment options were discussed with the patient. The patient was eventually fit with a left bilateral contralateral routing of the signal (BICROS) hearing aid.

54.1 Clinical History and Description

A 40-year-old female arrived at the clinic reporting a longstanding history of bilateral otosclerosis. A stapedotomy was performed in the right ear approximately ten years ago that provided some improvement in hearing. Although not stated, it is likely the right ear was selected because the operative report stated the right ear was the poorer ear. Unfortunately, the results from the audiological examinations before and after the stapedotomy were not available from the clinic where the stapedotomy was performed. The report from her six-month postoperative examination states “the audiogram done today reveals the air–bone gap on the right ear to have improved from about 40 to 60 dB HL to about 5 to 10 dB.” The left ear was described as having a severe to profound mixed hearing loss and the right ear to have a slightly sloping moderate sensorineural hearing loss (SNHL). Prior to being seen for the current audiological examination, the patient developed otitis media in her right ear resulting in a perforation. A few weeks later she experienced a sudden hearing loss, aural fullness, and tinnitus and extreme sensitivity to sound directed toward her right ear. She was placed on a high dose (the precise dose is unknown) of prednisone, acyclovir, and clindamycin, but the patient did not notice any improved hearing.

The patient arrived to the current audiology clinic for a second opinion for treatment of the right sudden hearing loss because her hearing did not improve with the use of oral steroids. The medical records of her previous audiograms before and after the sudden hearing loss were not available and were not placed in her medical chart. She had no prior experience with amplification, and she had significant difficulty communicating due to the hearing loss from otosclerosis in the left ear and sudden hearing loss in the right ear. Otoscopy revealed the tympanic membranes were intact bilaterally, indicating the perforation in the right eardrum had healed. The otologist performed a right transtympanic dexamethasone injection to determine if hearing could be improved in the right ear. The patient was loaned a Pocketalker® (Williams Sound, Eden Prairie, MN) so she could communicate with her spouse during medical management of the sudden hearing loss. An audiological examination and a follow-up appointment with the otologist were scheduled in 2 weeks.

54.2 Audiological Testing

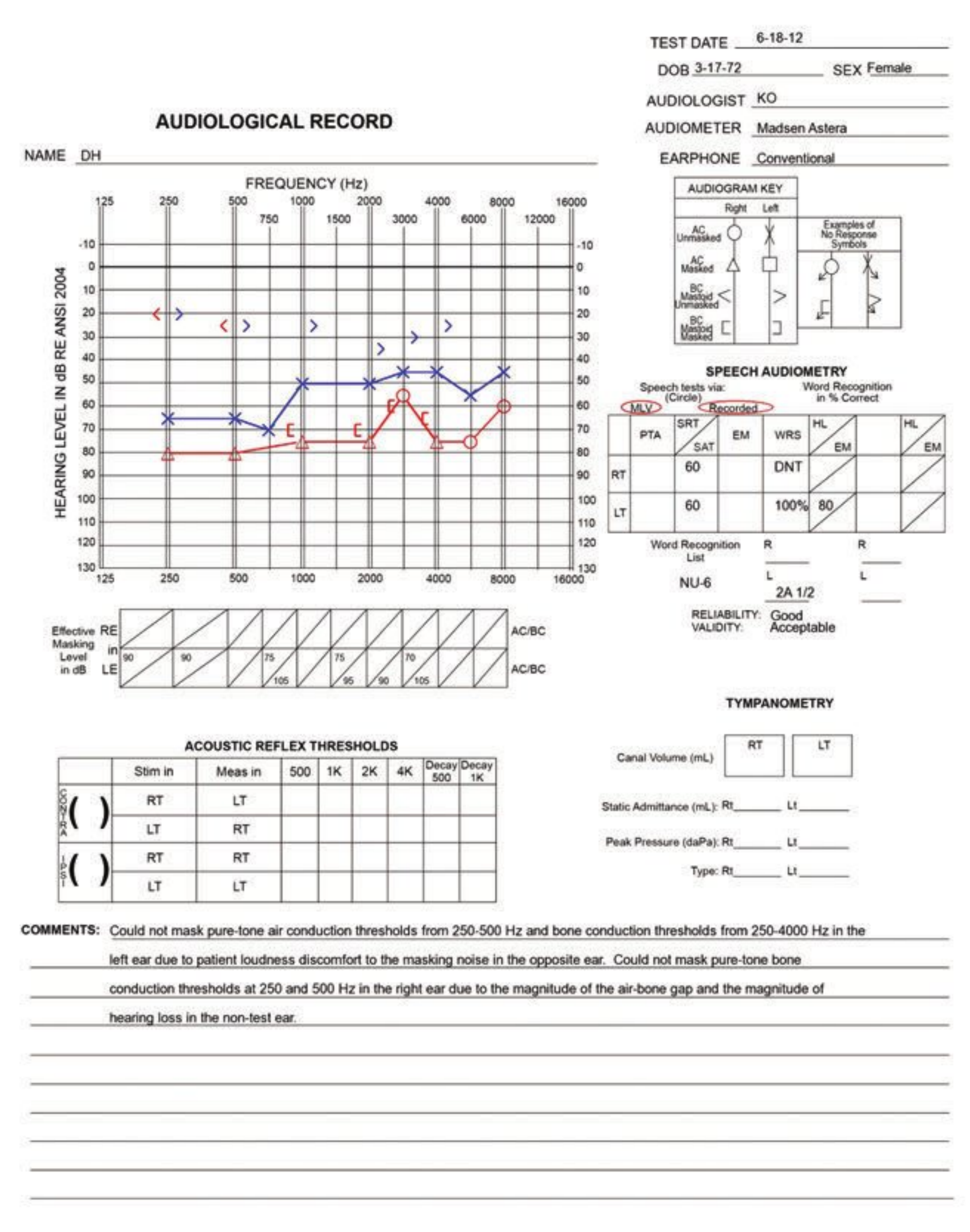
When the patient arrived for the audiological examination it was noted that an earplug was worn in the right ear due to the patient experiencing loudness discomfort to moderately loud sounds. Also, she reported she did not notice an improvement in hearing in the right ear. The results of the audiological examination two weeks after the first dexamethasone injection are reported in Fig. 54.1. The air-conduction thresholds at 250 to 500 Hz and bone-conduction thresholds at 250 to 4000 Hz could not be masked in the left ear due to patient loudness discomfort from the masking noise placed in the right ear. Also, bone-conduction thresholds at 250 to 500 Hz could not be masked in the right ear due to the air–bone gap and magnitude of hearing loss in the left ear. A speech awareness threshold (SAT) in the right ear revealed a moderately-severe loss in the ability to detect speech and a speech recognition threshold (SRT) in the left ear revealed a moderately-severe loss in the ability to receive speech. Word recognition scores (WRSs) using the Northwestern University Auditory Test Number 6 (NU-6) word lists with a recorded female talker revealed normal ability to recognize speech in the left ear and word recognition could not be measured in the right ear due to patient sensitivity to sound. Loudness discomfort levels (LDLs) were not performed due to limited time; however, it was noted that patient loudness discomfort was reported at only 10 dB sensation level (SL) relative to the air-conduction thresholds for narrow-band masking noise. Immittance testing was not performed due to the previous stapedotomy in the right ear. At this clinic, immittance testing on surgically modified ears is not performed unless requested by the otologist due to concern for disturbing/changing the surgical site.

54.3 Questions to the Reader

1. How would you describe the patient’s pure-tone thresholds in Fig. 54.1?
2. What amplification options could the patient use to communicate with her family and friends during the medical management of the right sudden hearing loss?

54.4 Discussion of Questions to the Reader

1. How would you describe the patient’s pure-tone thresholds in Fig. 54.1?
   The right ear presents with a severe mixed hearing loss at 250 to 2000 Hz, rising to a moderate SNHL at 3000 Hz, sloping to a severe SNHL from 4000 to 6000 Hz, and rising to a moderately-severe hearing loss at 8000 Hz. The left ear presents with a moderately-severe mixed hearing loss from 250 to 750 Hz that rises to a moderate mixed hearing loss at 1000 to 8000 Hz.
2. What amplification options could the patient use to communicate with her family and friends during the medical management of the right sudden hearing loss?
   One amplification option would be to fit the patient with a loaner hearing aid in the left ear. She is not a previous hearing aid user, however, and does not have a custom



TEST DATE 6-18-12
DOB 3-17-72 SEX Female
AUDIOLOGIST KO
AUDIOMETER Madsen Astera
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME DH

SPEECH AUDIOMETRY

Speech tests via: (Circle) [✓ MLV] [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		60		DNT			
LT		60		100%	80		

Word Recognition List R ___ R ___
NU-6 L 2A 1/2 L ___

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt_______ Lt_______
Peak Pressure (daPa): Rt_______ Lt_______
Type: Rt_______ Lt_______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: Could not mask pure-tone air conduction thresholds from 250-500 Hz and bone conduction thresholds from 250-4000 Hz in the left ear due to patient loudness discomfort to the masking noise in the opposite ear. Could not mask pure-tone bone conduction thresholds at 250 and 500 Hz in the right ear due to the magnitude of the air-bone gap and the magnitude of hearing loss in the non-test ear.

Fig. 54.1 Initial audiological examination.

earmold. Fitting her with a loaner earmold or comply tip, therefore, may be difficult due to discomfort as well as the possibility of feedback from the magnitude of amplification required for the degree of hearing loss in her left ear. Another option is providing hearing assistive technology (HAT), such as a Pocketalker, although the use of HAT is more cumbersome. Finally, she and her family members could be counseled on communication strategies, such as the following:

1. Face each other while communicating.
2. Speak in a slow and clear manner.
3. Be no greater than 3 feet from each other when communicating.
4. Sit in a quiet area of a restaurant.

54.5 Diagnosis and Recommended Treatment

Due to no noted improvement in hearing in the right ear, a second dexamethasone injection was performed and the patient was followed two weeks later. Again, the patient reported no significant improvement in hearing in the right ear, and a medical examination by the otologist did not reveal a cause for the sudden hearing loss. An audiological examination was performed and revealed no significant changes in the left ear and a decrease in hearing in the right ear by 10 dB HL at 500 and 3000 Hz and 25 dB HL at 6000 and 8000 Hz. The otologist decided not to perform further dexamethasone injections due to no noted improvement in hearing in the right ear. The otologist recommended the patient not pursue surgery on her left ear due to the risks involved. This risk included the possibility that the patient could lose further hearing in the left ear as a result of surgery, and because the left ear is her better ear, the patient agreed with the otologist's recommendation. The patient was medically cleared for amplification and referred for a hearing aid evaluation (HAE).

54.6 Additional Questions to the Reader

1. What are the realistic expectations from amplification that need to be addressed with this patient at the HAE because of the magnitude of hearing loss?

2. What recommendations for amplification would be appropriate?
3. Would fitting a hearing aid in the right ear be appropriate?
4. Would an auditory osseointegrated implant system (AOIS) be appropriate?
5. What are the advantages and disadvantages of a monaural hearing aid, BICROS, and a AOIS in the left ear?

54.7 Discussion of Additional Questions to the Reader

1. What are the realistic expectations from amplification that need to be addressed with this patient at the HAE because of the magnitude of hearing loss?
   Counseling should include a review of the audiogram and the anatomy and physiology of the ear, and discussion of the advantages and disadvantages of binaural versus monaural amplification based on the patient's magnitude of hearing loss. In the case of monaural amplification, options include a left monaural hearing aid, left BICROS, or left AOIS. The discussion concerning these monaural options should include the inability to restore localization, difficulty in background noise, and the effect of the head shadow effect.
2. What recommendations for amplification would be appropriate?
   The patient could benefit from a left monaural, left BICROS, left AOIS, or bilateral AOIS (which was suggested by the otologist).
3. Would fitting a hearing aid in the right ear be appropriate?
   Due to the patient's hypersensitivity to sound in the right ear, fitting a hearing aid to that ear did not seem to be an optimal option. If the hypersensitivity subsides, a hearing aid could potentially be used for sound awareness if this is a patient goal. LDLs could be performed to examine patient hypersensitivity in the future to determine if a hearing aid would be feasible.
4. Would an auditory osseointegrated implant system (AOIS) be appropriate?
   The U.S. Food and Drug Administration (FDA) criteria for patient selection for AOIS for patients with conductive or mixed hearing loss is a bone conduction pure-tone average of < 45 dB HL from 500 to 3000 Hz. Bilateral AOIS are intended for patients with moderate to severe symmetrical conductive or mixed hearing loss. Symmetric is defined as a difference no greater than 10 dB HL in the bone conduction pure-tone average of 500 to 4000 Hz or < 15 dB difference at individual frequencies 500 to 4000 Hz.
5. What are the advantages and disadvantages of a left monaural, left BICROS, left AOIS?
   a. A left monaural fit would allow the patient to hear signals from the left side, but the patient would continue to have difficulty hearing signals from the right side.
   b. A left BICROS would improve hearing in the left ear, eliminate the head shadow effect, and has been reported to provide objective and subjective benefit. The BICROS, however, has limitations in background noise, particularly when noise is on the transmitter (poorer side) side, because the omnidirectional microphone of the transmitter amplifies the noise and transmits the noise to the better ear, which may interfere with the wanted speech signal.
      A recent improvement in BICROS technology has incorporated a directional microphone on the receiver side (Phonak, U.S., Warrenville, IL) while the transmitter is active, which may help to improve speech recognition in noise. Unfortunately, Williams et al (2012) recently reported no significant differences between the participants' current omnidirectional microphone BICROS and the directional microphone BICROS when the words in noise (WIN) test was presented from the front and noise was presented from 180°, or speech was presented to the better ear and noise to the poorer ear, or finally, when speech was presented to the poorer ear and noise to the better ear.
   c. A left AOIS would improve speech recognition for signals originating from the left side, but the patient would still have difficulty hearing signals from the right side. Unfortunately, for this patient, her asymmetric hearing loss is outside the current FDA AOIS fitting criteria for bilateral AOIS. Currently, no research is available concerning the performance of bilateral AOIS for a patient with asymmetric hearing loss with unaidable mixed hearing loss in one ear and a mixed aidable ear on the opposite side. One study examined benefit with a AOIS implanted on the side with unaidable hearing and with a mild to moderate SNHL on the opposite ear and reported benefit even with the mild to moderate SNHL in the opposite ear. Although this study did not directly examine the implications of mixed hearing loss, the AOIS could potentially be an amplification option if the patient does not perceive benefit with the left monaural or left BICROS fit. An AOIS could be an amplification option based on her current hearing thresholds in the left ear; however, there is some concern about the possibility of a decrease in hearing in the left ear. If this were to occur, the AOIS may not be able to provide optimal benefit, and this concern must be discussed with the patient.

54.8 Outcome

The patient reported difficulty hearing family members, the television, in the car, and in background noise. At the HAE, different features and levels of the technology (directional microphones, bands/channels) were discussed as well as HAT to enhance the benefits from amplification, and the patient was extensively counseled on the possible advantages and disadvantages of the following:

1. Left monaural fit such as the improved hearing in the better ear, but the inability to localize sound, poor sound awareness from the right side, and difficulty in background noise
2. Left BICROS such as the ability to hear sounds on the right side in the left ear, but the inability to localize sound, and difficulty in background noise
3. A left AOIS would allow for speech recognition on the left side, but the patient would still experience difficulty hearing someone on the right side, localizing sound, and listening in background noise. A AOIS implanted to the right ear in conjunction with a left AOIS would allow sound awareness on the right side; however, the patient would not be able localize sound and would continue to have difficulty in background noise and she does not meet FDA criteria for bilateral implantation of AOIS.

During the HAE, the patient experienced a left Phonak Ambra microM behind-the-ear (BTE) hearing aid, left Phonak BICROS

(Ambra microM BTE receiver with a Phonak BTE CROS transmitter), and a left Cochlear Divino and the BP-100 AOIS connected to a headband. The Ambra microM and BICROS were preprogrammed by placing the patient’s audiogram into NOAH and downloading a “First-Fit.” For the AOIS, the default setting was downloaded, and the patient could increase or decrease the volume as needed. The Divino was used after demonstrating the BP-100 because patients usually report the gain at the factory setting for the BP-100 is “too soft.” After experiencing these amplification options, the patient reported the sound quality of the BP-100 sounded hollow and the Ambra microM and BICROS sounded tinny. The limitations of using the manufacturers’ “First-Fit” were discussed for the three devices. The patient was counseled that the device selected by the patient would be verified at the fitting using real-ear measures for the monaural and BICROS fitting or aided sound-field thresholds for the AOIS. The patient also inquired about a hearing aid for her right ear and was counseled that due to the hypersensitivity to sound in her right ear, a hearing aid was not recommended, but may be an option later if the patient is interested in sound awareness on the right side. If the patient reports a decrease in hypersensitivity in the right ear, LDLs as well as word recognition could be measured to determine if a hearing aid would be appropriate. Currently, however, an SL of 10 dB of masking noise when masking during audiometry was reported as uncomfortable, and WRS could not be performed due discomfort. These patient reports warranted a “wait and see” recommendation.

After extensive counseling, the patient decided to pursue a left Phonak Solana microP BTE BICROS receiver and an in-the-canal (ITC) transmitter. Further, she was counseled that if she did not perceive benefit with the BICROS, she could consider a monaural hearing aid or a trial with a AOIS on a headband to determine if these options provide better benefit and meet her communication needs.

Electroacoustic analysis of the hearing aid (ANSI S3.22–2003) to manufacturer specifications was obtained when the hearing aids arrived. Also, verification of transmission of the signal from the transmitter to the receiver and directional microphone performance was verified prior to the hearing aid fitting.

The patient was fit with the left BICROS using real ear insertion gain (REIG) measures at input levels of 50, 65, and 80 dB sound pressure level (SPL) using a speech-weighted composite signal so the measured REIG matched the National Acoustic Laboratories’ non-linear fitting formula version 1 (NAL-NL1) corrected for the air–bone gap and channel summation (16 channels). Loudness judgments were performed using a 50, 65, and 80 dB SPL speech-weighted composite signal, and soft sounds (50 dB SPL) were reported as “soft,” average input sounds (65 dB SPL) were reported as “comfortable,” and loud sounds (80 dB SPL) were reported as “loud, but okay.” The patient was counseled on how to use and care for the BICROS and returned in two weeks for follow-up.

At the follow-up, the patient reported she had gone to a concert and was pleased with the ability of the hearing aid to hear the music and the people around her. The patient also reported that she does well with the BICROS in quiet and when communicating on the telephone, but notices some difficulty in background noise and hearing the TV. To address these concerns, the mid and high frequencies of the “Speech in Noise” and “Calm” programs within the “Sound-flow” automatic program were increased by 2 dB to help improve speech recognition in these listening situations. The patient was reminded of the limitations of the BICROS in background noise. A Washington University School of Medicine Outcome Questionnaire was administered (Table 54.1) to determine aided benefit with the

Table 54.1 Responses at the patient’s hearing aid assessment using the Washington University School of Medicine Outcome Questionnaire.

Difficulty at home	Always	Often	Sometimes	Rarely	Never	N/A
Communicate with spouse	U		N			
Family members/friends	U		N			
Children	U		N			
TV, stereo	U		N			
Telephone	U		N			
Difficulty at work	Always	Often	Sometimes	Rarely	Never	N/A
Telephone	U		N			
One-on-one in noisy situations	U	N				
Small meetings	U			N		
Large meetings with speaker greater than 12 feet	U			N		
Difficulty in social situations	Always	Often	Sometimes	Rarely	Never	N/A
Family gatherings	U		N			
Noisy restaurant	U	N				
House of worship	U			N		
Theater	U			N		
Party	U	N				

Abbreviations: N, new; U, unaided. Source: Reprinted with permission from the Washington University School of Medicine.

BICROS ("N" for "New") compared to unaided ("U" for "Unaided") at home, work, and in social settings. The patient reported a slight improvement in background noise and significant improvement in quiet situations. The patient decided to keep the BICROS hearing aid and was provided with an electronic dehumidifier and counseled on how to use it. Two weeks later, the patient reported she was doing better in moderately noisy environments. The patient was provided with information to purchase a loud weather radio with a bed shaker to place near her bed due to the patient's concern of not hearing storms at night.

54.9 Key Points

1. It is important to counsel patients on realistic expectations of amplification for patients with aidable hearing in one ear and unaidable hearing in the opposite ear.
2. It is important to allow the patient to take an active role in deciding which amplification option is best and to incorporate HAT options into the counseling.
3. It is important to include outcome measures to determine if amplification has provided benefit for the patient in a variety of listening environments.
4. It is important to use a multidisciplinary approach when a patient presents with several hearing disorders to identify and treat each hearing disorder appropriately.

Suggested Reading

Del Dot J, Hickson LM, O'Connell B. Speech perception in noise with BICROS hearing aids. Scand Audiol 1992; 21: 261–264

Olsen SØ, Glad H, Nielsen LH. Comparison of two bone anchored hearing instruments: BP100 and Ponto Pro. Int J Audiol 2011; 50: 920–928

United States Food and Drug Administration. Section 2 summary and certification. 510(k) summary number K082108. 2008 http://www.accessdata.fda.gov/cdrh_docs/pdf8/k082108.pdf

Wazen JJ, Van Ess MJ, Alameda J, Ortega C, Modisett M, Pinsky K. The Baha system in patients with single-sided deafness and contralateral hearing loss. Otolaryngol Head Neck Surg 2010; 142: 554–559

Williams VA, McArdle RA, Chisolm TH. Subjective and objective outcomes from new BiCROS technology in a veteran sample. J Am Acad Audiol 2012; 23: 789–806

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55 A Case for Real Ear Measures

Kristi Oeding

BA is scheduled for a comprehensive audiological examination and hearing aid evaluation (HAE). He obtained bilateral receiver-in-the-canal (RIC) hearing aids 3 to 4 years ago from a large retail chain. He notices benefit with the hearing aids, but believes he is not hearing as well as he should, particularly in noisy listening situations. Real ear insertion gain (REIG) measures revealed minimal insertion gain bilaterally when compared with the prescribed National Acoustic Laboratories' non-linear fitting formula version 1 (NAL-NL1).

55.1 Clinical History and Description

BA is a 68-year-old male who was initially examined by an otologist due to BA reporting decreased hearing in the right ear following an upper respiratory tract infection that began two months ago. BA reported hearing loss since middle school due to an extensive history of ear infections as a child. BA also reported a number of myringotomies that included insertion of pressure-equalization tubes. BA stated that his right ear had better hearing until the upper respiratory tract infection. BA started wearing bilateral RIC hearing aids three to four years ago, and he stated he had a family history of hearing loss that included his grandfather. He had a significant history of noise exposure as a result of working in construction for approximately 40 years.

Otoscopy for the right ear revealed amber fluid behind the tympanic membrane and the tympanic membrane was retracted. Otoscopy for the left ear revealed that the tympanic membrane was also retracted, but fluid was not present behind the tympanic membrane. The otologist's examination reported right chronic otitis media, and BA's hearing loss was thought to be due to the combination of presbycusis, excessive exposure to noise, and genetics. The otologist recommended a follow-up appointment in one month with a repeat audiological examination and a head computed tomographic (CT) scan. The otologist also recommended using an Otovent (Abigo Medical AB, Askim, Sweden). The Otovent consists of a nose plug and latex balloon. The patient pinches one nostril shut and puts the Otovent in the other nostril and, by blowing air through the nose, inflates the latex balloon. Once the balloon is inflated, the air from the balloon reenters the nostril, creating positive pressure, the patient swallows and the eustachian tube opens allowing fluid to drain from the middle ear.

55.2 Audiological Testing

When BA returned in one month, he reported an immediate improvement in hearing in the right ear after using the Otovent. Along with an audiological examination, the patient was also scheduled for an HAE. Results of the audiological examination (Fig. 55.1) revealed hearing was within normal limits for the right ear from 250 to 1000 Hz and precipitously fell to a moderate to moderately-severe sensorineural hearing loss from 1500 to 8000 Hz. Results in the left ear revealed hearing to be within normal limits at 250 Hz and sloping to a slight mixed hearing loss from 500 to 1000 Hz, then precipitously falling to a moderate to moderately-severe sensorineural hearing loss from 1500 to 8000 Hz. Speech recognition thresholds (SRTs) were normal in the right ear and revealed a slight loss in the ability to receive speech in the left ear. Word recognition scores (WRSs) using the Northwestern University Test Number 6 (NU-6) word lists with a recorded female talker revealed normal ability to recognize speech bilaterally (92%bilaterally). Immittance audiometry was performed and revealed a normal tympanogram in the right ear and excessive negative pressure (− 105 daPa) in the left ear indicating the possible presence of a malfunctioning eustachian tube. Acoustic reflex thresholds (ARTs) for right ipsilateral stimulation were within normal limits from 500 to 2000 Hz and absent at 4000 Hz and for left ipsilateral stimulation were elevated at 1000 Hz, and absent at 500 and 2000 to 4000 Hz. ARTs for right contra lateral stimulation were elevated at 500 Hz and absent from 1000 to 4000 Hz and for left contralateral stimulation were within normal limits at 1000 Hz, elevated at 2000 Hz, and absent at 500 and 4000 Hz. Reflex decay could only be performed for left contralateral stimulation at 1000 Hz, because the right contralateral ART at 1000 Hz and left contralateral ART at 500 Hz were absent and the right contralateral ART at 500 Hz was elevated; therefore, the intensity of the stimulus could not be made loud enough (+10 dB SL) to perform reflex decay. The result of reflex decay for left contralateral at 1000 Hz was negative.

As mentioned earlier, BA wore bilateral RIC hearing aids with directional microphones and open domes. He purchased the hearing aids from a large chain, which was an approximately four hour drive away due to the low cost of the hearing aids compared with other clinics in his area. BA thought he received benefit from the hearing aids, but was disappointed with how he understands speech in noisy listening situations. BA questioned if his ability to understand speech in background noise could be improved.

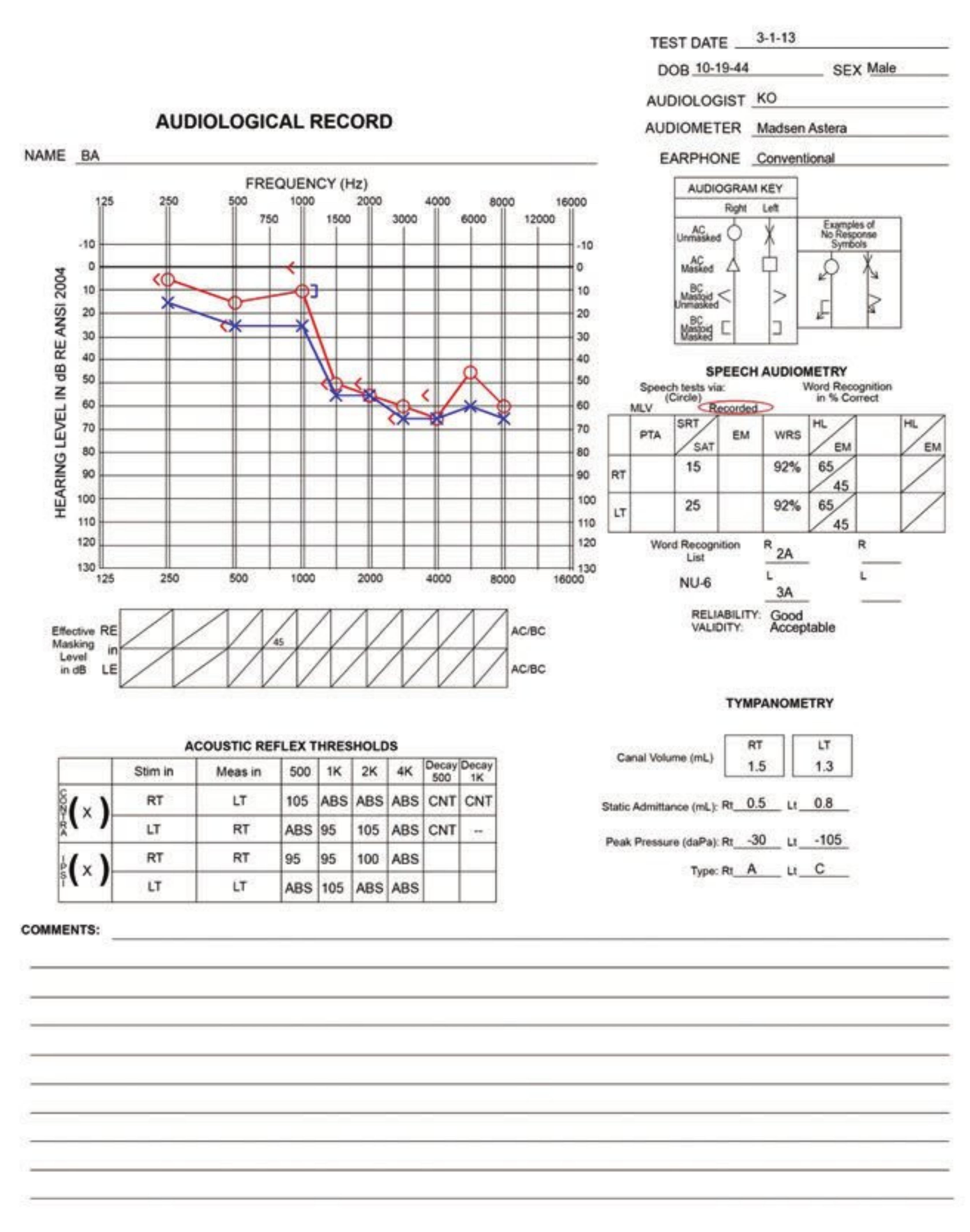
55.3 Questions to the Reader

1. What recommendations would a clinician provide BA about his hearing aids?
2. Are differences in gain/output to be expected between a manufacturer's "First-Fit" compared with using real ear measures (REMs) to program hearing aids?
3. What considerations need to be taken into account when performing REMs on an open-fit hearing aid?

55.4 Discussion of Questions to the Reader

1. What recommendations would a clinician provide BA about his hearing aids?

   The patient could be counseled about considering more current hearing aid technology or reprogramming his current



TEST DATE 3-1-13
DOB 10-19-44 SEX Male
AUDIOLOGIST KO
AUDIOMETER Madsen Astera
EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME BA

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		15		92%	65 / 45		
LT		25		92%	65 / 45		

Word Recognition List NU-6 R 2A L 3A R ___ L ___

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.5	1.3

Static Admittance (mL): Rt 0.5 Lt 0.8
Peak Pressure (daPa): Rt -30 Lt -105
Type: Rt A Lt C

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	105	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	95	105	ABS	CNT	--
IPSI (X)	RT	RT	95	95	100	ABS		
	LT	LT	ABS	105	ABS	ABS		

COMMENTS:

Fig. 55.1 BA's comprehensive audiological and immittance examination.

hearing aids. BA has had his hearing aids for three to four years and he may or may not notice improved benefit from more current hearing aid technology. Another option is to determine the amount of amplification being provided via REMs and to reprogram BA's hearing aids to improve benefit if the hearing aids are not providing adequate amplification. BA's hearing may also have changed since he purchased his current hearing aids, which could be a reason why BA is not hearing as well.

2. Are differences in gain/output to be expected between a manufacturer's "First-Fit" compared with using real ear measures (REMs) to program hearing aids?
   Several studies have reported significant differences between the manufacturer's First-Fit and a prescriptive fitting formula, such as NAL-NL1. For example, Aazh and Moore (2007) reported a difference of ± 10 dB from the NAL-NL1 prescriptive target at one or more frequencies between 250 and 4000 Hz using the manufacturer's First-Fit for 64% of ears that were measured. Another study by Abram et al reported higher benefit scores on the Abbreviated Profile of Hearing Aid Benefit (APHAB) and that more experienced hearing aid users preferred hearing aids programmed using REMs to NAL-NL1 than hearing aids fit according to the manufacturer's prescribed First-Fit. Also, a MarkeTrak report revealed that performing verification and validation during the hearing aid fitting process resulted in 1.2 fewer visits compared with not performing these measures. Verification refers to measures that confirm the hearing aid is working properly and fit appropriately for the patient (i.e., electroacoustic performance, REMs, etc.), and validation refers to measures that confirm the patient is obtaining/perceiving benefit from the hearing aid (i.e., aided speech in noise measures, questionnaires, etc.).
3. What considerations need to be taken into account when performing REMs on an open-fit hearing aid?
   One of the most important considerations is the use of the reference microphone. There are two methods to calibrate the loudspeaker of a REM system to ensure that signals (in dB sound pressure level [SPL]) from the loudspeaker to the reference microphone placed near the ear is at the stated input level when the signals reach the microphones of the hearing aid. One method is the modified pressure method with concurrent equalization. Using this method, the reference microphone is actively analyzing in real time the output

(dB SPL) of the loudspeaker to the reference microphone and adjusting the output based on patient movement so that the input level near the microphones of the hearing aid remains constant. The other method is the modified pressure method with stored equalization. Using this method, the reference microphone is only activated when the reference and probe microphones are leveled and when measuring the unaided response of the ear. Then the reference microphone is deactivated during aided measures, and it is critical that the patient hold his or her head still. The patient should be instructed to look straight ahead at the loudspeaker and to keep the head still for all measures; the audiologist must keep an eye on the patient to ensure this occurs.

When an open fitting is being verified in the aided condition, amplified sound could escape from the ear canal and reach the reference microphone. A confounding variable can occur with the modified pressure method with concurrent equalization because the amplified sound escaping the ear canal could reach the reference microphone causing the loudspeaker output to decrease due to the perceived higher output received by the reference microphone. This could result in the audiologist thinking there is less gain/output than is actually being provided by the hearing aid. The gain/output of the hearing aid would then be programmed to be increased to compensate for this in order to match the prescribed target. This could lead to overamplification of approximately 5 dB, particularly from approximately 1000 to 4000 Hz. Studies have reported that the concern of amplified sound interfering with the reference test microphone primarily occurred when the prescriptive target prescribed ≥25 dB of gain. Otherwise, ≤25 dB of gain had minimal impact on the reference microphone and the accuracy of the measured gain to target. Therefore, for open-fit hearing aid fittings requiring ≥25 dB of gain, the modified pressure method with stored equalization is recommended.

55.5 Diagnosis and Recommended Treatment

BA was counseled on reprogramming the hearing aids he purchased about three to four years ago or the possibility of obtaining new hearing aids. In this case, the hearing aids would have to be reprogrammed where the patient purchased his hearing aids due to the proprietary software required to program the hearing aids. If the software was not proprietary, but the audiologist did not have the software or cables to program the hearing aids, the audiologist could call the manufacturer to order the appropriate software and cables for the hearing aids to program them. Because the patient had the hearing aids for only three to four years, the option of verifying the magnitude of amplification the patient was receiving from his hearing aids using REIG measures in relation to the NAL-NL1 prescriptive fitting formula was discussed. If it was determined that the hearing aids were not providing appropriate amplification, the patient could use this information to return to the clinic where he purchased his hearing aids for reprogramming. A local office of the retail chain where the patient purchased his hearing aids recently opened close to his home, and the patient would not have to drive as far for reprogramming. BA was counseled that he would be charged a standard rate per every 15 minutes of REMs because he had purchased his hearing aids from another clinic. The patient decided he would like to briefly learn about new hearing aid technology and also decided to pay to have REMs performed on his hearing aids. The improvements in hearing aid technology since he purchased his current hearing aids along with hearing assistive technology (HAT) were discussed prior to REMs.

Prior to REMs, the hearing aids were dehumidified, and the wax guard and dome were examined to make sure each was free of wax. Electroacoustic analysis was performed using the Frye FONIX 6500® Hearing Aid Analyzer (Frye Electronics, Inc. ©, Tigard, OR) using ANSI S3.22–1996 and a completely-in-the-canal (CIC) coupler. Results revealed reference test gain of 20.5 dB in the right ear and 19.0 dB in the left ear, which showed that the patient's hearing aids were operating. The patient's audiological data were placed into the Frye FONIX 8000 Hearing Aid Analyzer®, and the NAL-NL1 target was corrected for binaural and channel summation (six channels). REIG measures were performed using a 65 dB SPL input level of the DigiSpeech (intermittent speech-weighted) signal (Fig. 55.2). Results revealed minimal gain bilaterally compared with NAL-NL1. One possible reason for the differences in coupler versus real ear gain may be due to the coupling of the hearing aid to the ear, because the hearing aid receiver had an open dome and loosely fit at the outside of his ear canal; this may allow the amplified sound to leak out of his ear canal. Another reason may be due to the acoustics of BA's ear canal (i.e., large ear canal, large real ear unaided gain [REUG], etc.) and the need for more gain to compensate for this. It was noted that BA had a large REUG (20 dB bilaterally around 2500 Hz), which means the hearing aid would need to provide more amplification to overcome the insertion loss of the hearing aid to meet the NAL-NL1 target, although it is expected that there would be minimal insertion loss with an open-fit hearing aid.

Although the reference microphone was not deactivated for testing, it is expected that having the reference microphone activated would not have resulted in poorer than measured REMs because the prescribed gain was less than 25 dB. The patient was counseled on the limited amount of amplification he is receiving with his hearing aids. He was provided with a copy of the results, as well as instructions on the frequency regions (1000 to 6000 Hz in the right ear and 1000 to 6000 Hz in the left ear) that needed more gain in order to provide appropriate amplification according to NAL-NL1.

55.6 Outcome

BA returned to the otologist two weeks later. It was noted that the CT scan was normal, except for slight fluid in the right mastoid. A medical examination revealed that BA no longer had retracted tympanic membranes or fluid behind his right eardrum. BA reported he was working with the dispensing professional at the large retail chain to have his hearing aids reprogrammed to a more appropriate level for his hearing loss.

55.7 Key Points

1. REMs are an important evidence-based tool that must be used to verify every hearing aid fitting.

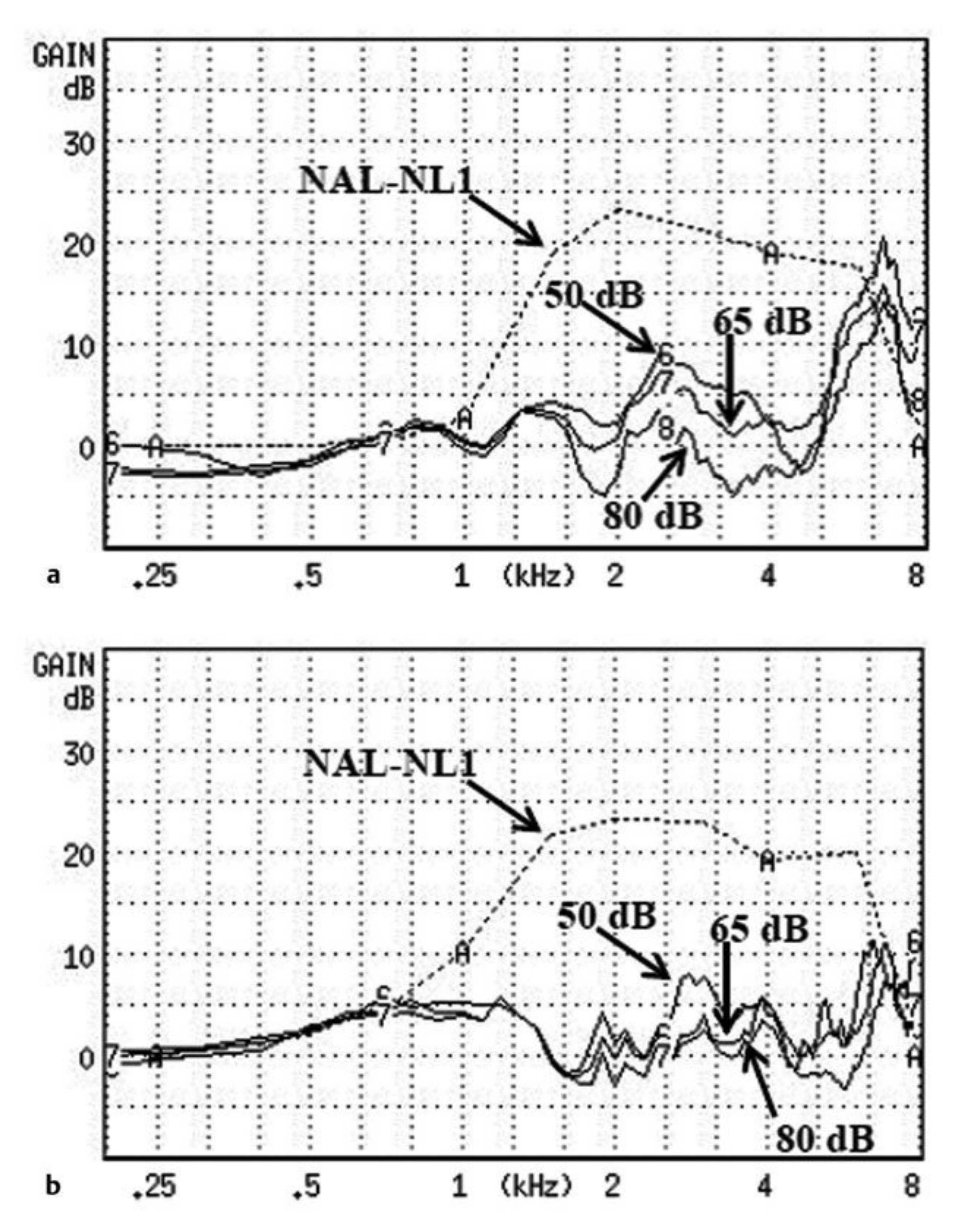


Fig. 55.2 Real ear insertion gain (REIG) measures for input levels of 50, 65, and 80 dB SPL using a DigiSpeech signal for the right (a) and left (b) ear. Also included is the NAL-NL1 target corrected for binaural and channel summation.

2. It is important to counsel patients with all available options so patients can make informed decisions about their hearing healthcare.
3. Audiologists should provide patients with information (i.e., performance of REMs, electroacoustic measures, charges for visits, etc.) on what to seek when selecting a facility to obtain hearing aids so patients will receive evidence-based practice care and maximum benefit from their hearing aid(s).

Suggested Reading

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56 Asymmetric Amplification Solution

Catherine V. Palmer

This case report describes an adult with moderate to profound mixed hearing loss in the right ear and profound hearing loss in the left ear. He is an active cyclist seeking a communication solution allowing him to receive input from both sides without compromising his ability to wear a bicycle helmet.

56.1 Clinical History and Description

AF, age 50, has what he describes as one “dead” and one “bad” ear. At age 16, AF underwent facial nerve decompression and mastoidectomy due to right facial paralysis secondary to acute suppurative otitis media and mastoiditis. At that time, the medical notes reported that AF had a congenital mandibular anomaly. A postoperative audiogram revealed hearing within normal limits for the left ear and a mild low-frequency conductive hearing loss in the right ear. Synkinesis (involuntary muscle movements following trauma to a nerve) was noted at the time of the postoperative audiological examination. Medical notes revealed that at age 20 AF underwent left stapes surgery with revision due to a perilymphatic fistula. At age 24, AF reports losing all hearing in his left ear during an airplane flight. An audiological examination from that time revealed air conduction thresholds in the left ear beyond the limits of the audiometer at 250 Hz and 8000 Hz and at 110 dB HL from 500 to 6000 Hz with no measurable bone conduction thresholds. No further medical treatment of the left ear was reported. Since age 30, audiological examinations have revealed a progressive, conductive hearing loss in the right ear of unknown etiology. The otologist’s notes revealed that exploratory surgery in the right ear was ruled out because of the left ear status (no usable hearing). The right ear currently has some exposed bone in the external auditory canal with a history of treatment for fungal infections and otitis media. Since age 40, AF has worn an in-the-canal (ITC) hearing aid in the right ear with no report of subsequent middle or outer ear infections.

AF owns and operates a successful retail business where he interacts with employees and customers throughout the day. AF indicates he misses important information if individuals speak from behind or from the left side. AF feels that on numerous occasions, customers have felt he was not interested or paying attention. When he is not interacting with people in person, he is on the phone with customers and suppliers to his business. AF indicates his current ITC hearing aid does not perform adequately with the telephone because it produces feedback when he places the phone close to his ear. Currently, AF removes his hearing aid when communicating on an amplified telephone. AF also reports attending noisy trade shows where he would like to have the ability to reduce the volume on his hearing aid. With his current ITC hearing aid, however, he does not have this capability, and he currently needs to remove the hearing aid. AF’s primary hobby is riding bicycles, and he rides alone or with a group. AF describes bicycle riding as essential to his mental and physical well-being and rides outdoors daily year-round. Currently, AF does not wear amplification when wearing his bicycle helmet due to feedback.

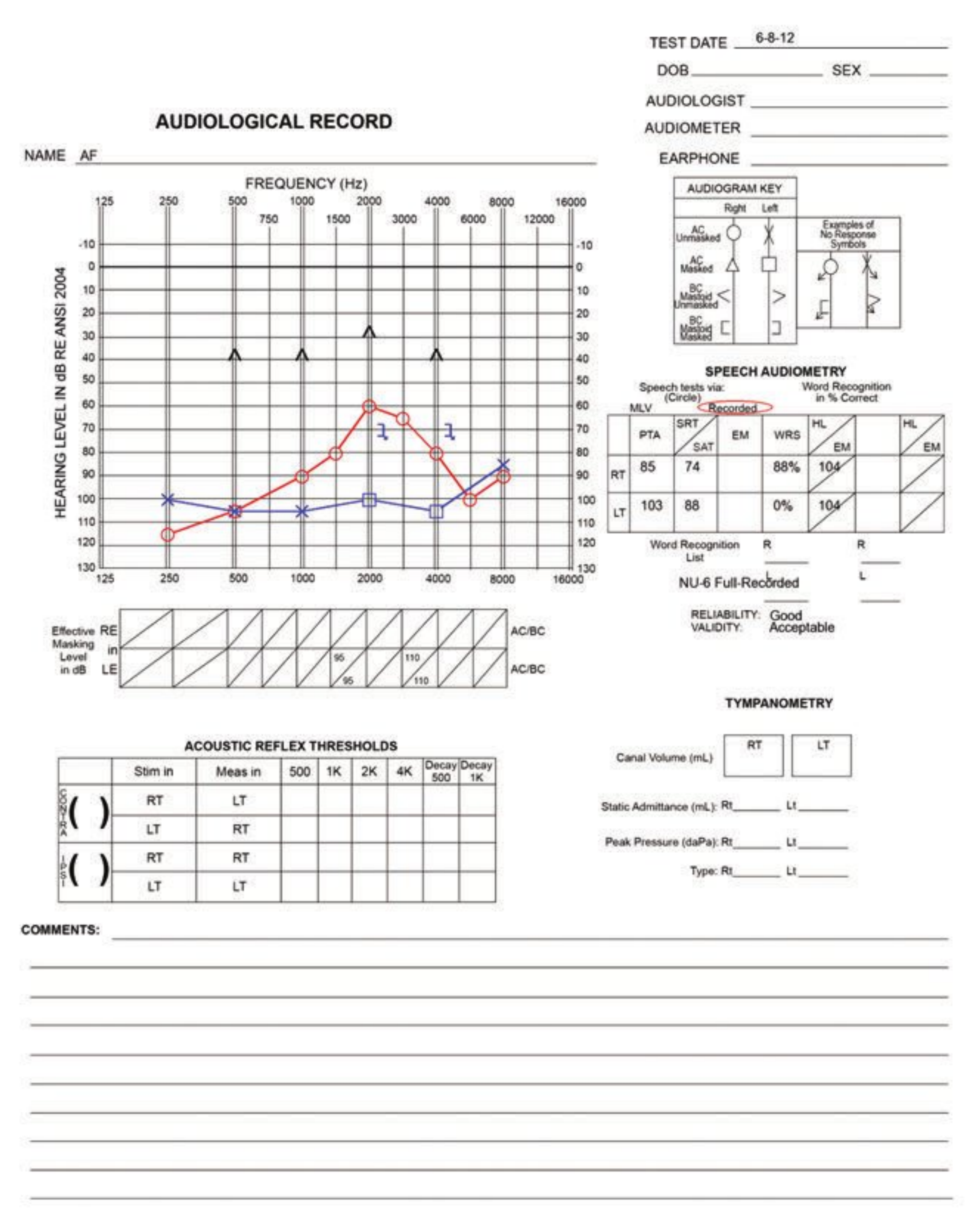
AF was referred to the audiology clinic by his otologist. AF visits the otologist every 6 months to ensure his middle and outer ears are healthy and free of fungal or bacterial infection. At his last visit, AF indicated to the otologist that his current amplification was no longer adequate for his communication needs. At this time, the otologist indicated that AF was not a candidate for further reconstructive surgery, but he was a candidate for an auditory osseointegrated implant system (AOIS). AF indicated that he did not want any type of surgery, including the implantation of an AOIS. Therefore, the otologist referred AF to the audiology clinic to discuss nonsurgical treatment options.

56.2 Audiological Testing

The results of AF’s most recent audiological examination are reported in Fig. 56.1. The left ear reveals a flat, profound hearing loss at 250 to 8000 Hz, which is presumed to be primarily sensorineural with no measurable bone conduction thresholds. Air and bone conduction thresholds in the right ear did not allow for sufficient masking to determine threshold in the left ear at 1000 Hz. Clearly, AF has a profound hearing loss in his left ear, but it may be poorer than what was recorded at this frequency. The right ear revealed a profound mixed hearing loss at 250 to 1000 Hz recovering to a moderate mixed hearing loss at 1500 to 3000 Hz and decreasing to a profound mixed hearing loss at 4000 to 8000 Hz. The pure-tone average (PTA) and speech recognition threshold (SRT) are in good agreement for the right ear. A speech awareness threshold (SAT) was obtained for the left ear because AF could not understand the words in order to repeat them. Word recognition scores (WRSs) at 104 dB HL revealed slight difficulty in the ability to recognize speech in the right ear and very poor ability to recognize speech in the left ear. AF completed an expectation worksheet where he listed what he would expect from a new amplification solution. These goals included hearing customers with less effort and more accuracy, hearing effectively on the telephone without removing his hearing aid, communicating effectively at employee meetings when people are seated on both sides, and being able to hear while riding his bicycle.

56.3 Questions to the Reader

1. Ideally, what are the goal(s) of a hearing aid fitting for AF?
2. What barriers are there to provide adequate amplification for AF?
3. What special considerations in AF’s lifestyle need to be taken into account when creating communication solutions?
4. What hearing aid options would be appropriate for AF based on his hearing loss and communication goals?



AUDIOLOGICAL RECORD

NAME AF

TEST DATE 6-8-12
DOB SEX
AUDIOLOGIST
AUDIOMETER
EARPHONE

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	85	74		88%	104		
LT	103	88		0%	104		

Word Recognition List R L R L

NU-6 Full-Recorded

RELIABILITY: Good
VALIDITY: Acceptable

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

TYMPANOMETRY

Canal Volume (mL) RT LT
Static Admittance (mL): Rt_____ Lt_____
Peak Pressure (daPa): Rt_____ Lt_____
Type: Rt_____ Lt_____

COMMENTS:

Fig. 56.1 Results from AF's current audiological examination.

56.4 Discussion of Questions to the Reader

1. Ideally, what are the goal(s) of a hearing aid fitting for AF?
   The general goals of the hearing aid fitting would include reception of sound originating from both sides of AF without the need for AF to reposition himself. Other goals would be improved recognition of speech when listening on the telephone and no feedback when wearing a bicycle helmet. Further, the amplification system should not promote chronic ear disease.
2. What barriers are there to provide adequate amplification for AF?
   AF's hearing loss is a potential barrier to adequate amplification. His profound left sensorineural hearing loss precludes the use of conventional amplification in that ear. The severity of the hearing loss in the right ear has limited the hearing aid style options that can be used due to feedback that is typical with smaller-sized instruments. The size/style of the amplification system is an issue because AF is an avid bicycle rider and has indicated this activity is essential to him. For safety reasons, he needs to hear during this activity. Previously reported chronic outer and middle ear disease may be a barrier for inserting a deeply seated, largely unvented instrument that will most likely be required to avoid feedback. The progressive nature of the hearing loss in the right ear may suggest that a solution that meets AF's needs today may not be effective in the future. This may require purchasing a new amplification system in the future.
3. What special considerations in AF's lifestyle need to be taken into account when creating communication solutions?
   AF's demanding work environment, which includes communicating in a busy retail business and meeting with employees, must be considered in any amplification solution. In addition, AF describes his bicycle riding as essential to his well-being and indicates he needs to hear during this activity

for safety. This latter requirement also creates special considerations in terms of configuration and style of the amplification solution.

4. What hearing aid options would be appropriate for AF based on his hearing loss and communication goals?
   Because the primary goal for AF is to perceive sound regardless of the side of the sound source, the amplification system must take sound received on the left side and deliver that sound to the less impaired right side. This is because the left ear is unable to make use of sound delivered directly based on the profound nature of the hearing loss. By definition, this means that a system that routes sound from the left ear to the right cochlea is needed as well as conventional amplification in the right ear. There are several options available for this combination of routing of signals, but some will not be consistent with the patient's goals for his new amplification system. A bilateral contralateral routing of signal (BICROS) system uses a microphone positioned on a behind-the-ear (BTE), in-the-ear (ITE), or completely in-the-canal (CIC) style hearing aid to receive sound from the left side. A transmitter from that hearing aid sends the sound to a right hearing aid that contains a receiver as well as a conventional amplification system for the right ear. This can be accomplished using a wireless system. AF's communication goals could be met with this amplification system in daily communication encounters. AF may or may not be satisfied with this system when using the telephone (land-line and cellular) because of the need to position the telephone specifically around the hearing aid. This solution may be prone to feedback when AF wears a bicycle helmet because of the microphone location on either a BTE or ITE hearing aid.

   A conventional bone conduction hearing aid (worn on a band to ensure adequate pressure to the left mastoid or seated into the ear canal attached to a BTE aid) could be used on the left side to receive sound and transmit it through vibration of the skull to the right cochlea. In this case, AF would also be fit with a conventional air conduction hearing aid in the right ear. It is unclear if sufficient power could be provided to the bone vibrator to elicit a response in the right cochlea because of reduced sensory bone conduction thresholds in the right ear (approximately 30 dB HL). An AOIS on the left mastoid would produce a more efficient transfer of energy and would be more likely to produce vibrations that could be received by the right cochlea. In either bone conduction option, the device would not be able to be worn comfortably under a bicycle helmet. In the case of the AOIS, AF made it clear that he did not want to undergo further surgery.

   A newly developed a device proposed for unilateral hearing loss (UHL) involves dental prosthesis that receives input from a microphone worn over the poorer ear using a BTE style device. The signal is converted into vibration, which is applied to the teeth and thereby transmitted via bone conduction to the functioning right cochlea. AF's right bone conduction thresholds (~ 30 dB HL) are at the limit for this device. Additionally, his medical report indicates that he has a congenital mandibular anomaly, which, upon further inspection, reveals dentition that will not support this device.

   Given the severe to profound nature of the hearing loss bilaterally, use of a cochlear implant (CI) might be appropriate. In AF's case, his mid- to high-frequency hearing sensitivity in the right ear is too good for him to be considered a CI candidate. Currently, audiometric thresholds poorer than 70 dB HL bilaterally are indicated for CI implantation. In addition, individuals who receive benefit from hearing aids are not considered appropriate candidates for a CI.

   A transcranial contralateral routing of the signal (CROS) hearing aid applied to the ear canal of the left ear and accompanied by conventional air conduction amplification in the right ear may be an amplification system that will meet all of AF's needs and provide AF with a successful communication solution in a variety of settings. A transcranial CROS fitting consists of a high-output air conduction hearing aid that is programmed to deliver sufficient amplified sound (i.e., output) via air conduction and cross over to reach the bone conduction thresholds of the opposite (better) cochlea. Successful users tend to have profound hearing loss in the affected side, so the high output of the air conduction amplification, which often sounds distorted is not heard by the significantly impaired cochlea. A transcranial CROS system is easiest to implement if the better ear has normal bone conduction thresholds because it would take less output to stimulate the cochlea at audible levels. In AF's case, he has no measurable bone conduction results in the left ear. This bodes well for forwarding the signal to the right cochlea without stimulating the very damaged left cochlea. Also, in AF's case, the better ear is quite impaired with a mild loss via bone conduction. This suggests that greater output from the aid worn in the left ear will be required to stimulate hearing in the right cochlea. Currently, audiologists are able to implement this solution with CIC hearing aids because of an increase in power now available in the CIC and as a result of improved feedback reduction strategies allowing for greater output prior to feedback. Even with advanced feedback strategies, it is essential that a deep, tight fit be achieved in order to obtain the required output for the left and right hearing aids for AF. If the left CIC transcranial CROS hearing aid can be programmed to allow the appropriate output to transmit sound to the right cochlea and a right CIC conventional hearing aid can be programmed with sufficient gain to allow soft (55 dB sound pressure level [SPL]), moderate (65 dB SPL), and loud (75 dB SPL) sounds to be audible, then all of AF's goals can be achieved through this configuration. If successful, AF will receive sounds from both sides, and he will be able to use the telephone while holding the phone naturally, as well as be able to wear his bicycle helmet when using this amplification configuration.

56.5 Additional Questions to the Reader

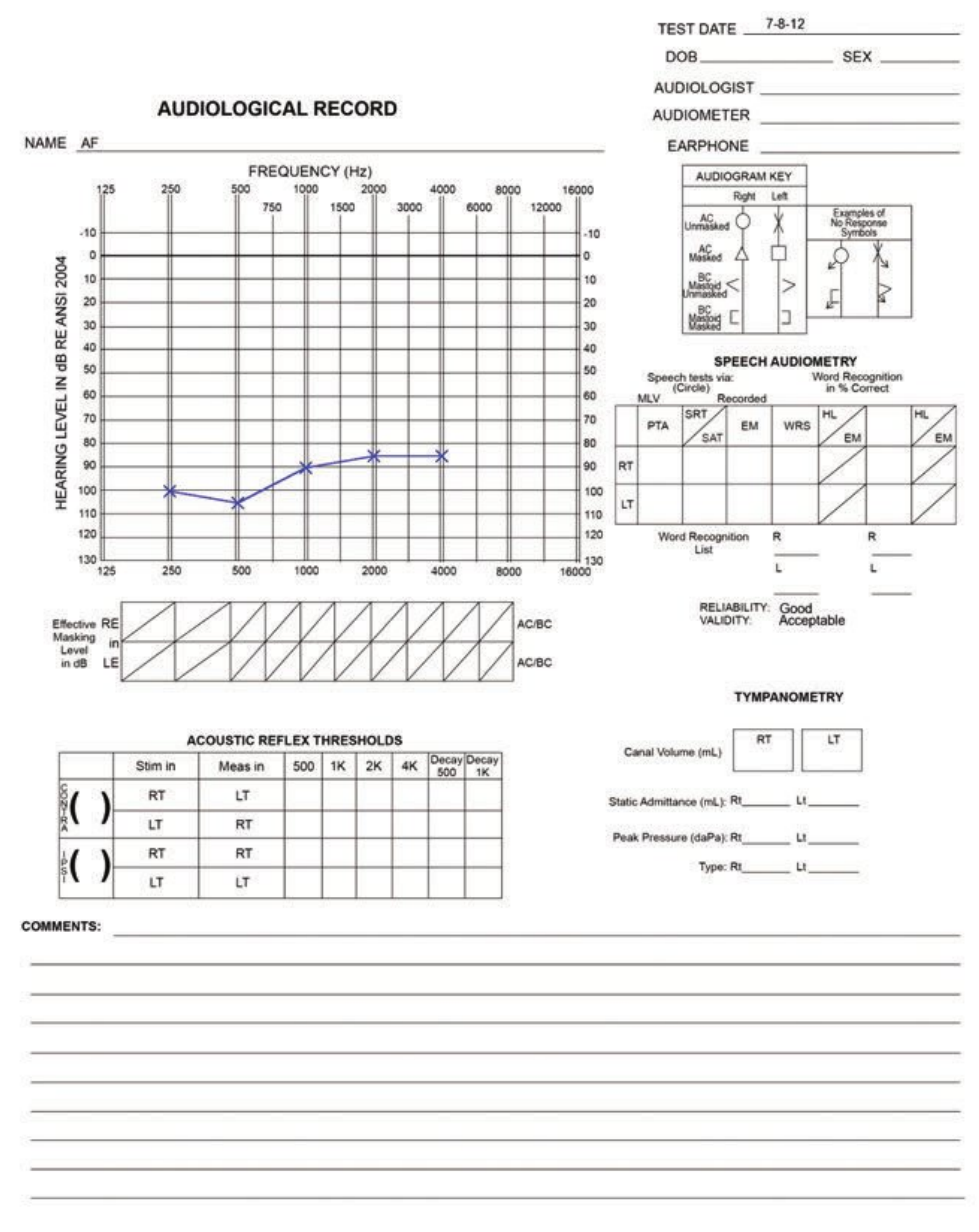
1. How will the transcranial CROS instrumentation be programmed to ensure crossover to the cochlea of the right ear via bone conduction with the use of the least possible output?
2. How will the right hearing aid be fit to ensure audibility of incoming sounds?
3. What solution could be offered to allow AF to manipulate the volume of the amplification in particularly noisy situations (e.g., trade shows)?

56.6 Discussion of Additional Questions to the Reader

1. How will the transcranial CROS instrumentation be programmed to ensure crossover to the cochlea of the right ear via bone conduction with the use of the least possible output?

   Once the hearing aids arrived and AF was ready to be fit, he was seated in a sound-treated room to evaluate the hearing thresholds of his left ear. At this visit, the hearing thresholds were measured without masking in the right ear. This indicates the lowest level of sound across frequency that can be applied to the left ear and to elicit a response in the right cochlea (Fig. 56.2). These thresholds were entered into the probe microphone system after applying the real ear to coupler difference (RECD) to accurately transform the dB HL into dB SPL to generate the target thresholds. Fig. 56.3 reports the real ear aided response (REAR in dB SPL) for the left ear. A probe microphone was inserted into the left ear and the left CIC was placed into the ear canal. The hearing aid software was adjusted until the output for a soft input level (---) delivered to the hearing aid from a loudspeaker arrived as close to the target (X-X) as possible. The "+" symbols represent the prescriptive target for this hearing loss, but the goal was to have the output for the soft input level arrive as close to threshold (X-X) as possible. The "*" symbol represents the uncomfortable loudness level (UCL). The measured REAR for the soft input level ("---") reported in Fig. 56.3 should be audible in AF's right cochlea.
2. How will the right hearing aid be fit to ensure audibility of incoming sounds?

   The aided output response for the right CIC was also verified by measuring the RECD and applying the RECD to transform the dB HL thresholds to dB SPL thresholds in the probe microphone system (as already explained for the left ear). A probe microphone was placed in the right ear canal with the CIC hearing aid. (See Fig. 56.4 for the results of the REAR testing.) Soft (55 dB SPL,---), moderate (65 dB SPL,___), and loud (75 dB SPL,—) sounds were delivered to the hearing aid from a loudspeaker, and the resulting output is displayed against AF's dynamic range and desired sensation level (DSL) [i/o]5.0 targets (++). The measured REAR for the input levels of 55–65–75 dB SPL met the prescriptive target at 500 Hz to



TEST DATE 7-8-12

DOB

SEX

AUDIOLOGIST

AUDIOMETER

EARPHONE

AUDIOLOGICAL RECORD

NAME AF

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT							
LT							

Word Recognition List R L R L

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt Lt

Peak Pressure (daPa): Rt Lt

Type: Rt Lt

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 56.2 Left unmasked thresholds.

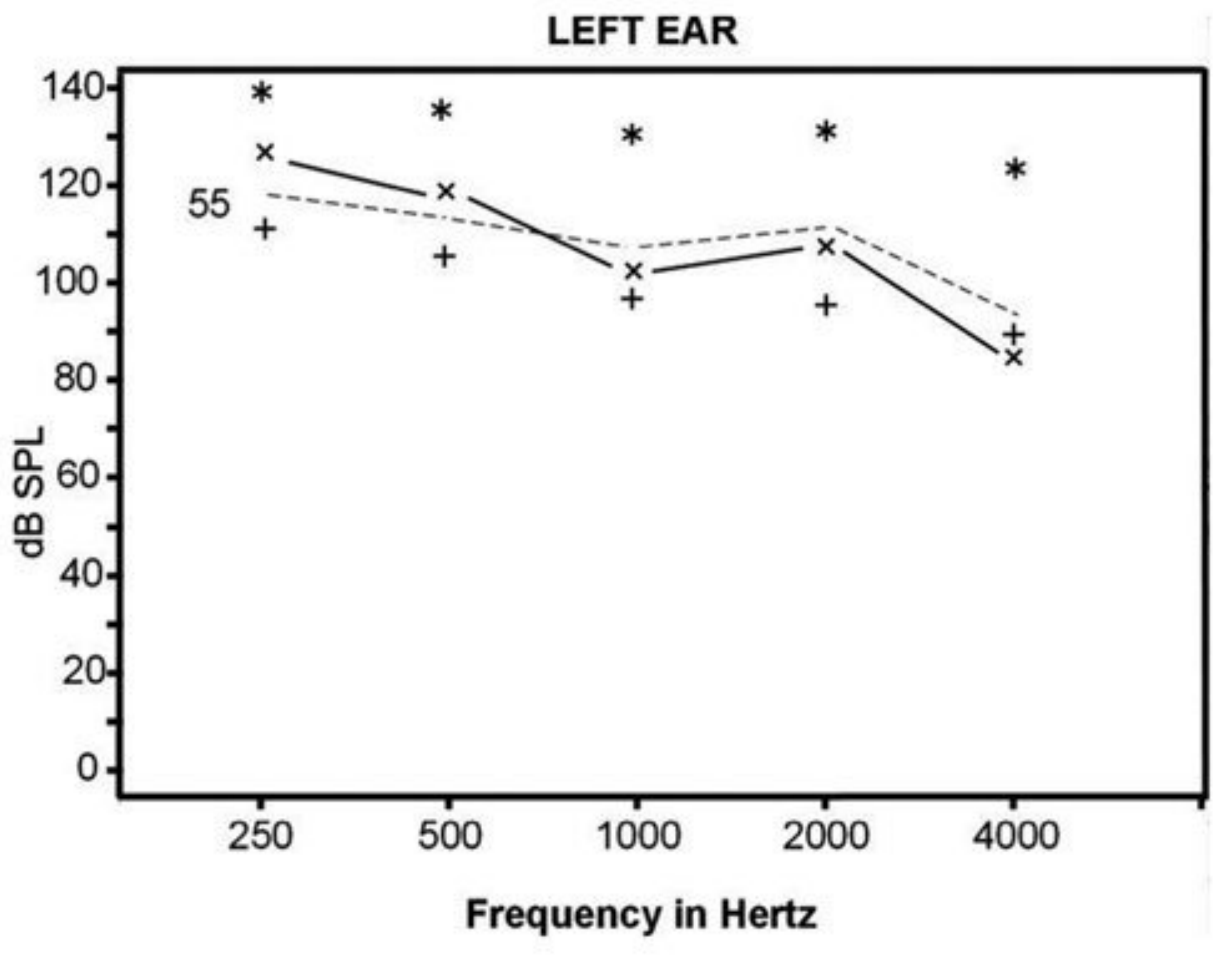


Fig. 56.3 Real ear aided response (REAR in dB SPL) probe microphone measures verifying that the output delivered to the left ear would be received by the right cochlea at an audible level. X, threshold; +, prescriptive targets; *, uncomfortable loudness level; ---, measured output for a 55 dB SPL input.

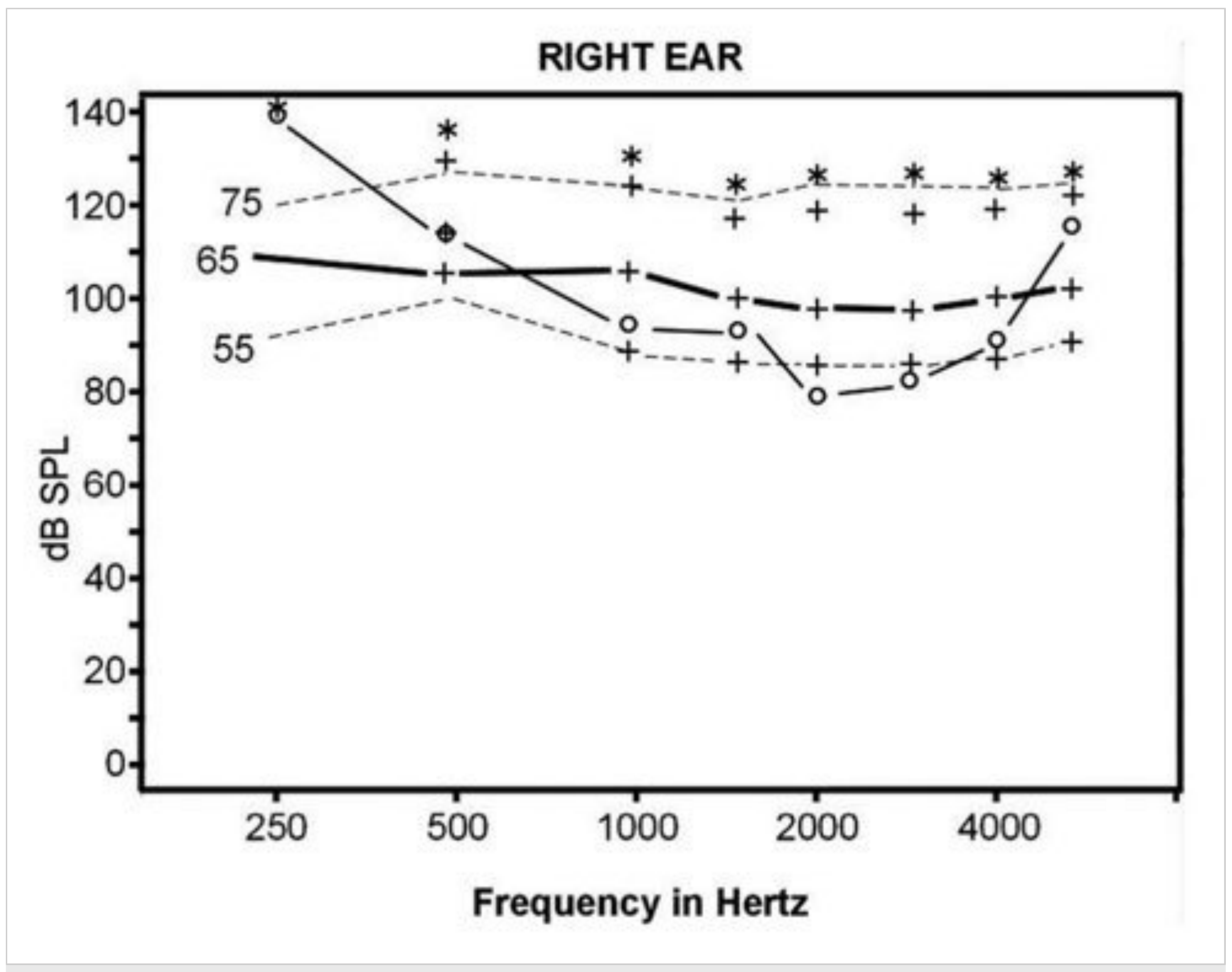


Fig. 56.4 Real Ear Aided Response (REAR in dB SPL) probe microphone measures verifying that soft, moderate, and loud input signals are audible to the right ear. (O = threshold, + = prescriptive target, * = uncomfortable loudness level,---= measured output for a 55 dB SPL input, ___ = measured output for a 65 dB SPL input,---= measured output for a 75 dB SPL input)

4000 Hz, suggesting that audibility was achieved for AF across a reasonably broad frequency range. Audibility was not obtained at 250 Hz due to the severity of the hearing loss as indicated by the measured REAR being below threshold at 250 Hz. Overall, the goal for audibility for AF was achieved.

3. What solution could be offered to allow AF to manipulate the volume of the amplification in particularly noisy situations (e.g., trade shows).

   A push-button volume control (VC) was ordered for each CIC so AF would be able to control the overall output of the hearing aids. For the left ear, AF may want to increase or decrease the output based on his perception of how he is receiving sound from the left side. AF also may use this VC in situations where noise is primarily arriving from the left side. The VC on the right CIC was ordered so AF would be more comfortable in noisy situations. This could have been accomplished by ordering a remote control, but AF did not want to carry a remote control to accomplish changes to his hearing aids.

56.7 Outcome

The CIC transcranial CROS instrumentation to the left ear, combined with the CIC conventional air conduction hearing aid to the right ear, provided AF with a solution that met his expectations and allowed AF to successfully communicate at work and on the telephone. Further, AF is able to hear while bicycle riding. AF reports using the VC to provide listening comfort in specific, noisy listening situations.

56.8 Key Points

1. A detailed case history, communication abilities, and needs assessment are essential in developing an appropriate amplification solution for patients.
2. Carefully defining the goal of the amplification system, in this case transferring sound to the better ear while amplifying the better ear, allowed the audiologist to consider all viable options and identify the option that best matched AF’s lifestyle requirements.
3. Real ear probe microphone measures are an essential tool in adjusting the appropriate output for nontraditional amplification devices.

Suggested Reading

Valente M. Fitting options for unilateral hearing loss. Hear J 1995; 48: 10, 45–48
Valente M, Oeding K. Transcranial contralateral routing of the signal as a fitting option for patients with single-sided deafness. Semin Hear 2010; 31: 366–377

57 Unilateral Hearing Loss and a Contralateral Routing of the Signal Fitting

John Pumford

LD is a 45 year old female being seen for a CROS evaluation and fitting.

57.1 Clinical History and Description

LD is a 45-year-old woman presenting with right unilateral hearing loss (UHL) following a head injury occurring as a result of an automobile accident. She was referred by a local physician to determine which intervention options may be suitable to address LD's listening difficulties. LD reports there are occasions where she manages just fine, but she is becoming increasingly frustrated by her inability to hear sounds originating on her right (poorer ear) side. LD mentions the hearing in her left ear is normal and she manages to "get by" in quiet listening environments when she can position her left ear toward sounds she is interested in hearing. Larger group situations, however, present greater difficulty, particularly when she is unable to turn her left ear toward signals of interest. In general, LD also finds it tiring to continually turn her better ear toward sounds she wants to hear and finds she sometimes misses sounds because she cannot orient herself quickly enough. Localization is also a concern as she finds it more difficult to locate the origin of sounds. Conversation in the car is reported as difficult, particularly when she is driving because her right ear is facing the inside of the automobile. LD has a demanding position at work where she is often asked to participate in and lead group meetings. She indicates occasional difficulty hearing those around the conference table, particularly when the speakers are on her right side. Discussion with the audiologist identified a number of potential intervention options for LD, including true transcranial contralateral routing of the signal (CROS) (i.e., bone-anchored hearing solution [BAHS]), quasi-transcranial CROS (i.e., deep-fitting completely in the canal [CIC]), and a conventional CROS hearing instrument (wired or wireless). LD was not interested in a surgical solution and expressed concerns with potential medical complications and the overall aesthetics of a BAHS. The provision of a quasi-transcranial solution was ruled out given LD's reported right ear canal sensitivity (i.e., occasional otalgia) following her automobile accident. LD and the audiologist determined that a conventional wireless CROS system was the desired solution. Fitting and verification of the selected CROS system was conducted to ensure appropriate performance and maximize benefit.

57.2 Audiological Testing

Otoscopic examination was unremarkable bilaterally. Pure-tone air and bone conduction audiometry was consistent with normal hearing on the left ear and a profound sensorineural hearing loss on the right ear (Fig. 57.1). Tympanometry revealed normal tympanic membrane performance bilaterally. Speech recognition thresholds (SRTs) were consistent with pure-tone results and revealed normal hearing for the left ear and a profound hearing loss for the right ear. Word recognition scores (WRSs) conducted at a most comfortable listening level revealed normal ability to recognize speech for the left ear and could not be conducted with the right ear given the magnitude of hearing loss.

57.3 Questions to the Reader

1. What type of listening difficulties might a patient with UHL experience?
2. What type of hearing instrument technology would be appropriate for a patient with UHL?
3. What type of coupling options are appropriate for conventional CROS/bilateral contralateral routing of signal (BICROS)?
4. What are the verification steps when fitting conventional CROS?
5. What are the expected benefits of CROS?

57.4 Discussion of Questions to the Reader

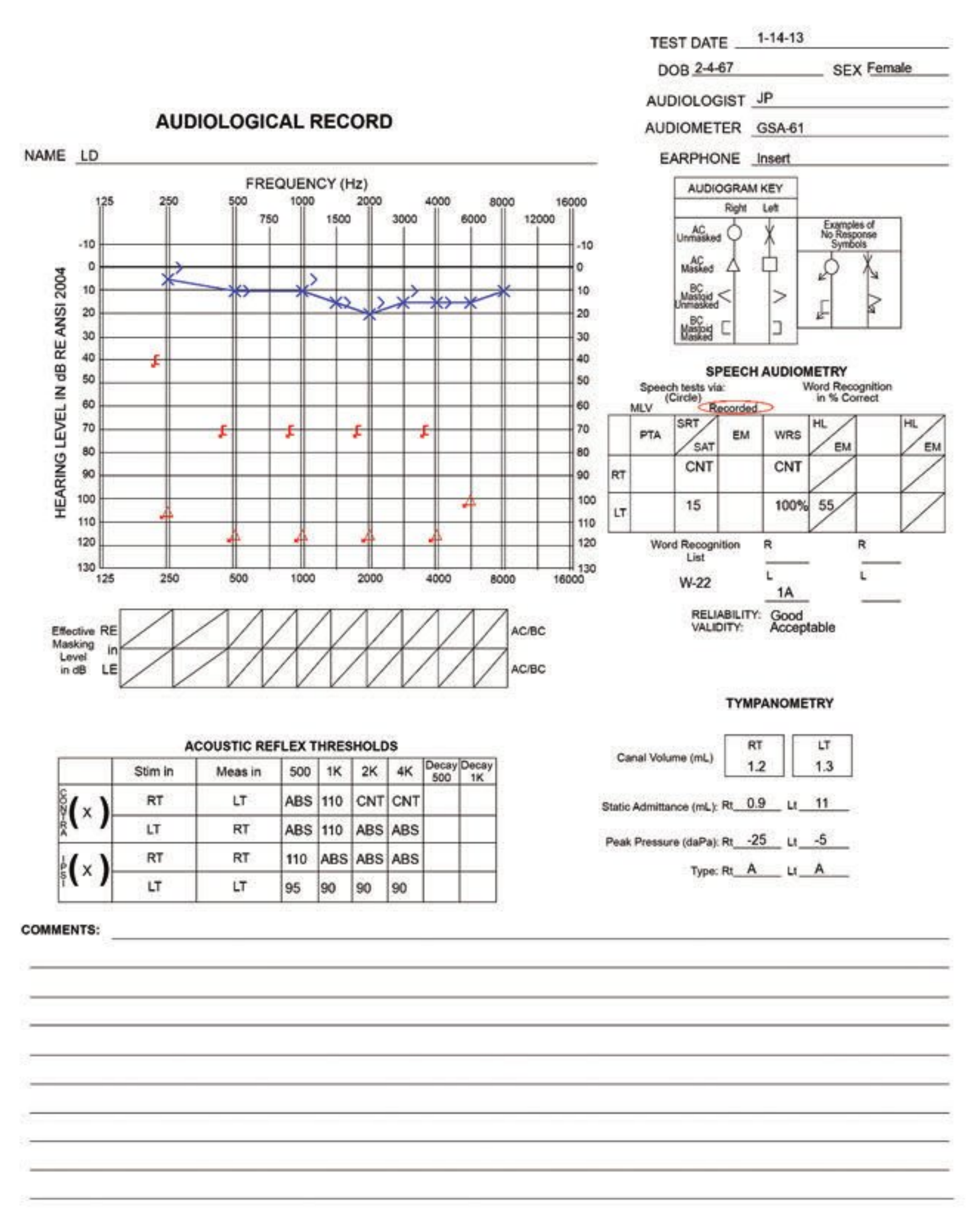
1. What type of listening difficulties might a patient with UHL experience?

   Patients such as LD who present with UHL are unable to take advantage of the benefits of binaural hearing, including, but not limited to, reduction of the head shadow effect, the squelch effect, sound localization, and binaural summation.

   a) Head shadow effect: The head casts an acoustic shadow and reduces the intensity level of sound received by the ear on the side of the head opposite the sound source. This phenomenon will result in a significant negative impact on LD's speech recognition for signals originating on the side of her poorer ear.

   b) Squelch effect: This term refers to the use of binaural cues to reduce the impact of background noise on speech recognition. Markides reported that individuals could reduce the detrimental effects of background noise and improve the ability to attend to a desired signal when the central auditory nervous system had the ability to analyze differences in time of arrival, phase, or intensity of signals between the two ears. Given LD's hearing loss, she will be unable to make use of the binaural squelch effect and will likely have greater difficulty listening in noise than those who have access to binaural cues.

   c) Localization: Due to the lack of binaural input to the central auditory nervous system, individuals with UHL typically report significant difficulty localizing sound in their environment. LD's central auditory nervous system is no longer able to compare acoustic aspects such as the time



TEST DATE 1-14-13

DOB 2-4-67 SEX Female

AUDIOLOGIST JP

AUDIOMETER GSA-61

EARPHONE Insert

AUDIOLOGICAL RECORD

NAME LD

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		CNT		CNT			
LT		15		100%	55		

Word Recognition List W-22

R

L 1A

R

L

RELIABILITY: Good

VALIDITY: Acceptable

Effective Masking Level in dB	RE										AC/BC
	LE										AC/BC

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.2	1.3

Static Admittance (mL): Rt 0.9 Lt 11

Peak Pressure (daPa): Rt -25 Lt -5

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	ABS	110	CNT	CNT		
	LT	RT	ABS	110	ABS	ABS		
IPSI (X)	RT	RT	110	ABS	ABS	ABS		
	LT	LT	95	90	90	90		

COMMENTS:

Fig. 57.1 LD's audiological examination.

of arrival and intensity differences of signals between ears to assist her in determining the location of the sound source.

d) Binaural summation: This phenomenon, where a signal received bilaterally is perceived as louder than the same signal received monaurally has been shown to result in an improvement in detection of sound. One would expect LD to notice a general reduction in her ability to detect sounds given her inability to benefit from binaural summation.

2. What type of hearing instrument technology would be appropriate for a patient with UHL?

Candidacy for different CROS amplification options for patients with UHL is based on the magnitude of residual hearing in the better ear. Patients with unaidable UHL having normal to near normal hearing in their better ear would be considered CROS candidates, whereas patients with some magnitude of hearing loss in their better ear would be considered better candidates for BICROS. CROS devices place a microphone (transmitter) on (or over) the unaidable ear that sends the signal to the better ear. As described by Valente et al, there are different approaches for transferring the signal. In general, these approaches can be divided into three primary categories:

a) Conventional: The signal is transferred to the better ear via an air conduction path (e.g., CROS or BICROS).
b) Quasi-transcranial: The signal is delivered via a bone conduction path using an air conduction technique (e.g., deep-fitting CIC).
c) True transcranial: The signal is delivered via the bone conduction path using a bone conduction technique (e.g., BAHS; SoundBite, Sonitus Medical, San Mateo, CA).

A full discussion of the quasi-transcranial and true transcranial systems is beyond the scope of this case report. Readers interested in learning more about these systems can refer to Valente et al and Dillon.

With the conventional CROS approach selected for LD, signals from the transmitter device are forwarded to the left ear via a wired or wireless approach. Given the normal hearing on the left ear side, the microphone on the receiving unit on the left ear is inactive. The goal is to transfer sound from the right ear side to overcome the difficulties associated with the head shadow effect. Like the CROS, the BICROS also places a microphone (e.g., transmitter) on the unaidable ear, which forwards signals originating on that side to the better ear. In contrast to the CROS system, however, given the hearing loss on the better ear side, amplification of sound originating on the side of the better ear is now required. As a result, to ensure audibility of sound all around the patient, a second microphone is placed over (in) the better ear. Sounds from both microphones are combined and processed by the hearing aid before being delivered to the better ear. Given LD's hearing loss with unaidable hearing in her poorer right ear and normal hearing in her better left ear, she was deemed to be a candidate for CROS, and a conventional wireless system meeting her needs was selected.

3. What type of coupling options are appropriate for conventional CROS/BICROS?

With conventional CROS and BICROS, various coupling options for the poorer and better ear sides are available. In CROS and BICROS, the key consideration for the poorer ear is appropriate retention of the transmitter device on the ear because sound is not delivered to that ear. Depending on the manufacturer and system selected, audiologists' choices for retention of the transmitter device may include a custom earmold, retainer earhook, or slim-tube. The form of the transmitter device may vary as well. The transmitter style may be a behind-the-ear (BTE) or in-the-ear (ITE) style. LD was fitted with a BTE wireless transmitter device on the poorer ear side with retention provided by a slim-tube attachment.

When considering appropriate sound delivery coupling options, one needs to consider the magnitude of hearing on the better ear. Given the normal hearing on the better ear of LD, it is important that the audiologist does not occlude the ear canal of the better ear. Leaving the ear canal open allows sound originating on the better ear side to pass unimpeded to the tympanic membrane. As such, an open/nonoccluding sound delivery option is appropriate. Depending on the system being fitted, this open fitting can be provided by a slim-tube with an open dome or a nonoccluding earmold (e.g., CROS mold, free-field earmold). LD was fitted with a slim-tube with open dome to ensure the better ear was not occluded. Should LD have had hearing loss in her better ear (i.e., BICROS candidate), a coupling/vent configuration suitable for the hearing loss on that ear would have been selected.

4. What are the verification steps when fitting conventional CROS?

When conducting real ear measurements (REMs) with conventional CROS, there are several principles to remember as outlined by Dillon and Pumford. First, the probe tube is always located in the ear canal of the better ear because this ear is receiving the amplified signal. Second, to ensure appropriate input signal levels during REMs, the reference microphone used to measure and control the input signal from the REM loudspeaker must be on the same side as the loudspeaker. If the REM system does not allow separation of the probe microphone and reference microphone, disabling the reference microphone via the software and using a stored equalization calibration method is preferable. Third, the loudspeaker is moved within a range of ± 45° relative to the front of the patient depending on the stage of the fitting process.

With CROS, as described earlier, the goal of the fitting is not to amplify sound but rather to transfer sound from the unaidable ear to the better ear to minimize the negative impact of the head shadow effect. As such, the audiologist's goal is to adjust the output of the receiver unit to match the ear canal resonance of the better ear (i.e., real ear unaided gain [REUG] or real ear unaided response [REUR]) such that the system sounds acoustically transparent to the patient. Use of the appropriate open-style coupling as described earlier ensures that sound originating on the better-ear side will pass through the ear canal unimpeded and be heard naturally with little to no occlusion. LD's CROS was fit and verified using REMs. The first step in this process is obtaining the open ear response for the better ear side as follows:

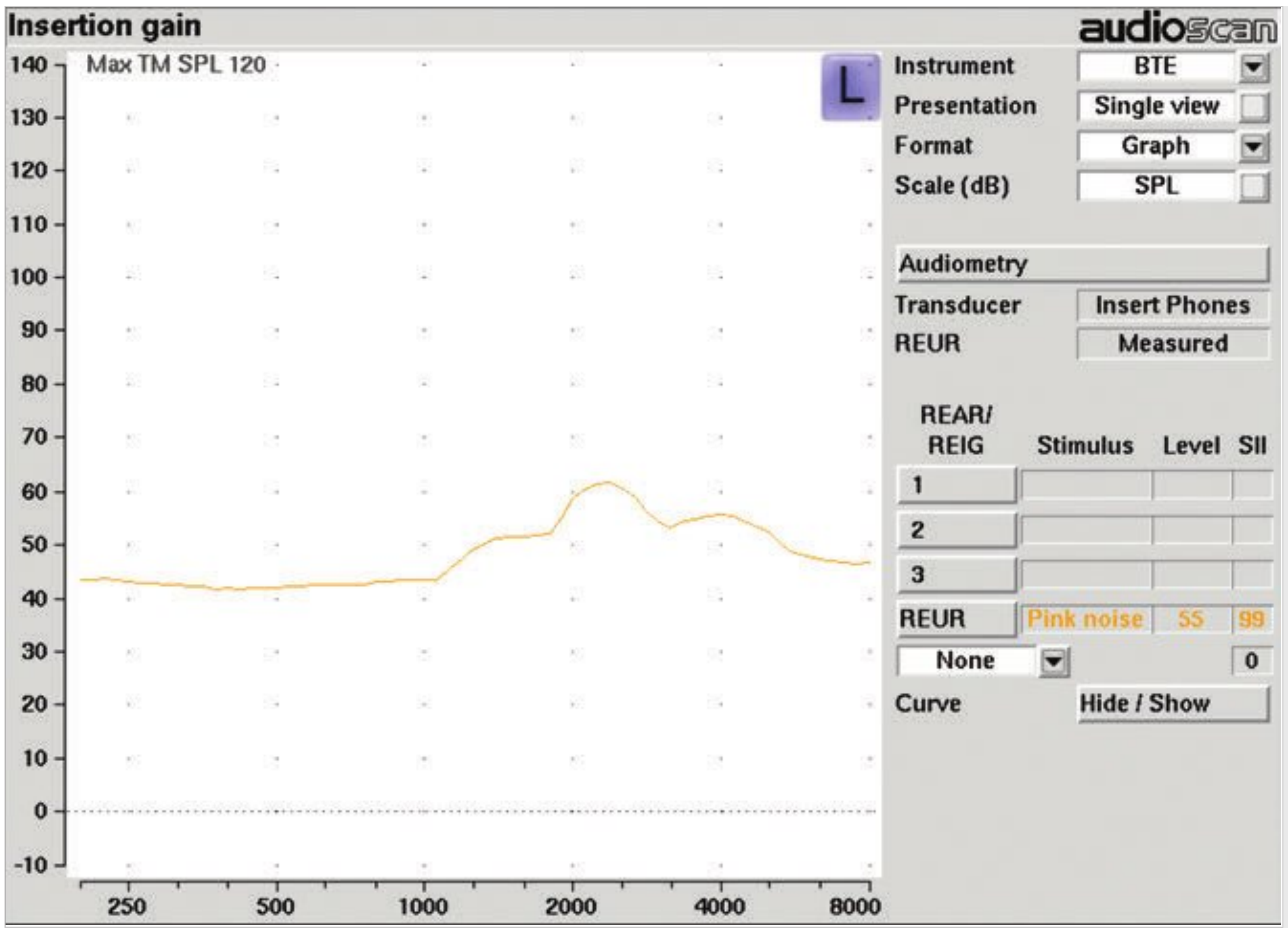
a) Position the reference microphone at the patient's better ear.
b) Insert the probe tube into the better ear.
c) Place the CROS BTE transmitter and BTE receiver units on the patient's right and left ears and turn these devices on.
d) Rotate the patient by 45° so the better ear is facing the sound source.
e) Record the REUR for the selected input signal (e.g., 55 dB sound pressure level [SPL] pink noise).

As reported in □ Fig. 57.2, this process resulted in a typical REUR for LD's better ear and verified that the open dome was nonoccluding. Note: if desired, the foregoing process could have been conducted initially with the open ear canal (no earpiece in place) to provide an "open ear" reference for step 1. Next, the second step is measuring the response for signals presented to the poorer ear as follows:

f) Rotate the patient so that the poorer ear is facing the loudspeaker (i.e., 45° azimuth).
g) Position the reference microphone at the poorer ear (same side as loudspeaker).
h) Leave the probe tube in the better ear.
i) Leave the CROS instruments on the ears and turned on as in step 1.
j) Record the real ear aided response (REAR) using the same input signal as in step 1. Note: When using a steady noise signal for verification, it is important to deactivate the noise reduction (NR) to avoid contamination of the measurement (i.e., reduction of device output due to activation of NR).

5. What are the expected benefits of CROS?

The primary benefit expected for patients fitted with CROS technology is increased awareness of sound occurring on their unaidable side (i.e., reduction of the head shadow effect). It follows that there may be improved speech recognition in situations where speech is directed toward the poorer ear. Unfortunately, the use of an amplification system that transfers sound to only one ear will not improve auditory functions that require binaural cues. As a result, localization and speech recognition in noise typically continue to be problematic, and patients should be counseled to supplement these concerns using other communication and amplification strategies (e.g., FM systems, visual cues, strategic positioning relative to signals of interest).



Insertion gain

Max TM SPL 120

Instrument	BTE
Presentation	Single view
Format	Graph
Scale (dB)	SPL

Audiometry

Transducer	Insert Phones
REUR	Measured

REAR/ REIG	Stimulus	Level	SII
1			
2			
3			
REUR	Pink noise	55	99
None			0

Curve Hide / Show

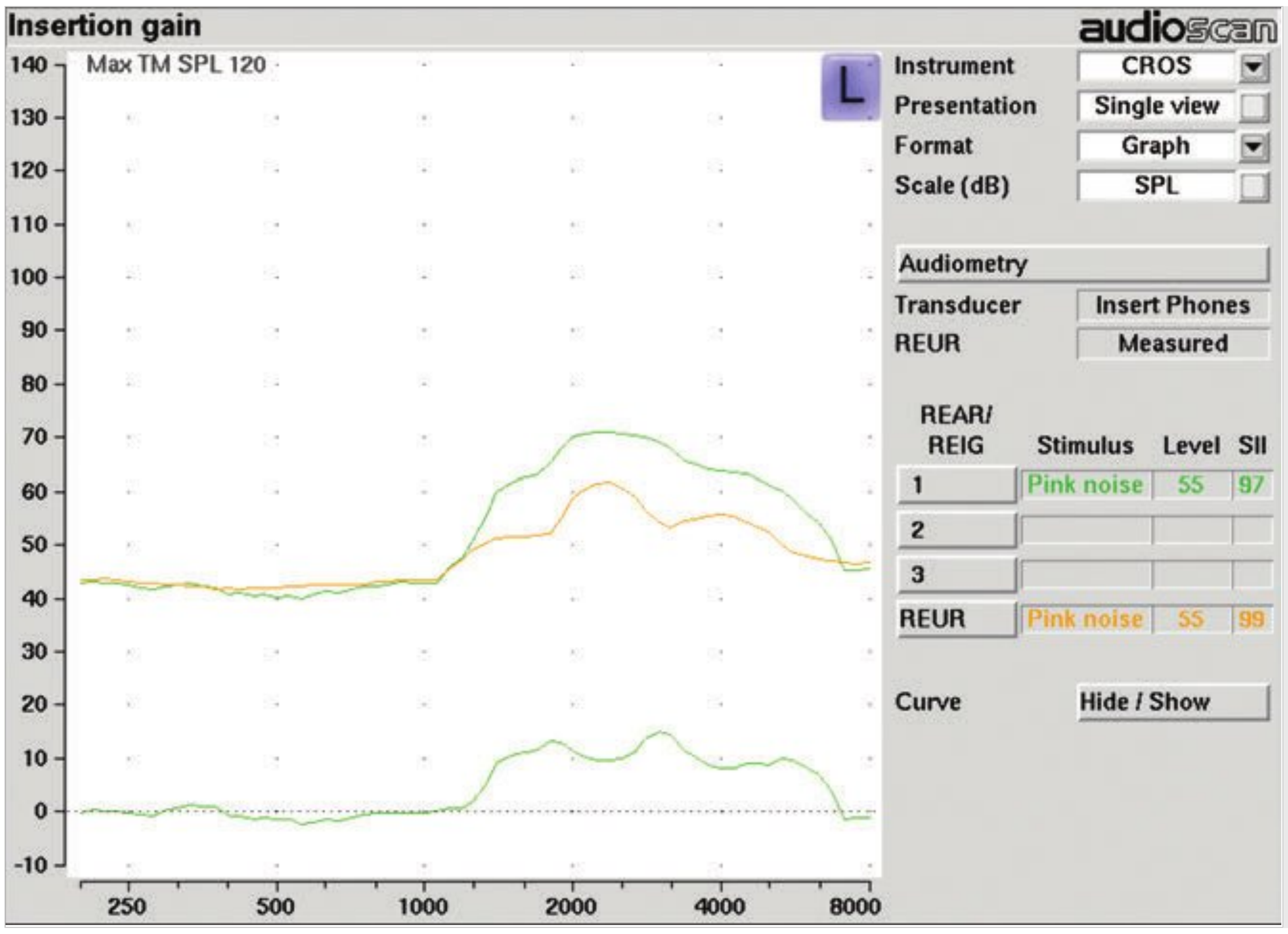
Fig. 57.2 Step 1: Measurement of LD's better ear side response.

57.5 Diagnosis and Recommended Treatment

Following the preceding verification steps, the initial fit of the CROS revealed overamplification of sound originating on the side of the poorer ear. As can be seen in Fig. 57.3, the initial quick fit of the CROS exceeded the initial REUR from ~ 1500 to 6000 Hz by ~ 10 dB. Initial comments from LD with the initial fitting in response to her own voice and the audiologist's voice were that the CROS sounded "too tinny" and "metallic." Subsequent adjustment of the gain/frequency response of the CROS as observed in Fig. 57.4 resulted in a closer match to the REUR and reports of improved sound quality by LD.

57.6 Outcome

Following her 30-day trial, LD reported benefit from her CROS due to her increased ability to hear sounds occurring all around her. Listening in her car became less frustrating because of her improved ability to hear others to her right while driving. Localization of sound, however, continued to present difficulties. Although LD had greater awareness of sound, she reported being unable to locate its origin based upon hearing alone and needed to visually scan the environment around her. LD also reported occasional difficulty using the CROS in noisy environments. In some instances LD found more difficulty than before (e.g., if nondesired signals were on her poorer ear side).



Insertion gain

Max TM SPL 120

Instrument	CROS
Presentation	Single view
Format	Graph
Scale (dB)	SPL

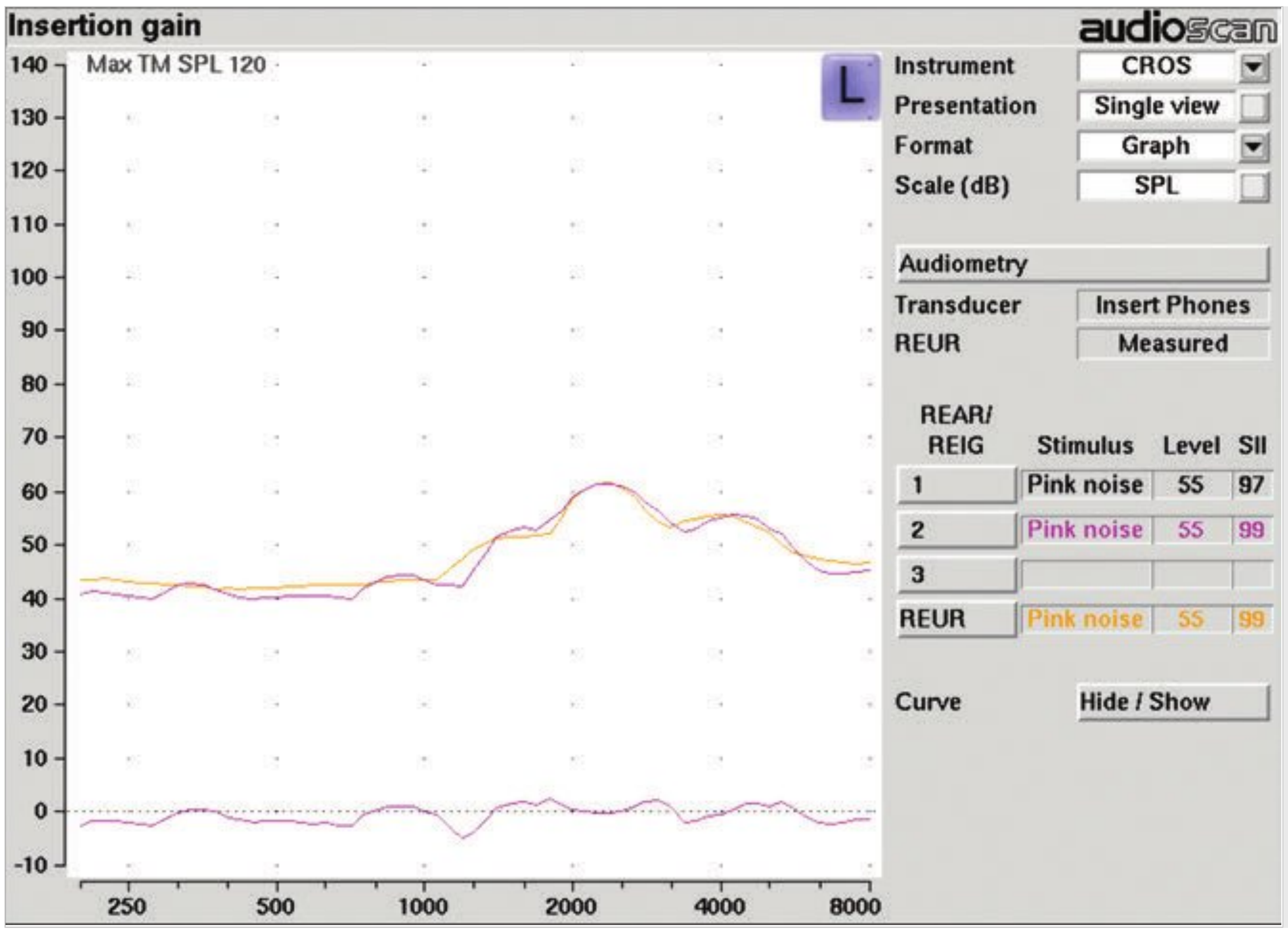
Audiometry

Transducer	Insert Phones
REUR	Measured

REAR/ REIG	Stimulus	Level	SII
1	Pink noise	55	97
2			
3			
REUR	Pink noise	55	99

Curve Hide / Show

Fig. 57.3 Step 2: Initial measurement of LD's poorer ear side response.



Instrument: CROS
Presentation: Single view
Format: Graph
Scale (dB): SPL

Audiometry
Transducer: Insert Phones
REUR: Measured

REAR/ REIG	Stimulus	Level	SII
1	Pink noise	55	97
2	Pink noise	55	99
3			
REUR	Pink noise	55	99

Curve: Hide / Show

Fig. 57.4 Step 2 Measurement following adjustment of LD's poorer ear side response.

Counseling included discussion of the value of reducing the gain of the transmitter by reducing the volume control or deactivating the transmitter in these types of listening situations, such that the better ear can actually benefit from the head shadow effect. Overall, LD reported she found sufficient benefit with the CROS to continue using it in situations where she was unable to turn her ear toward the signal of interest.

57.7 Key Points

1. UHL can create complex listening difficulties due to the patient's loss of binaural cues.
2. Various amplification options are available for those with UHL, subdivided by the method by which signals originating on the side of the poorer ear are received by the better ear.
3. The level of residual hearing in the better ear determines whether a patient is a CROS (normal hearing in better ear) or a BICROS (hearing loss in better ear) candidate.
4. Conventional CROS requires an open/unoccluded coupling option for the patient's better ear to ensure that sound originating on that side is received naturally and sound quality issues related to occlusion are not created.
5. Conventional BICROS requires a coupling option for the patient's better ear that is suitable for the hearing loss present on that ear.
6. REM is a valuable tool when fitting CROS or BICROS.
7. To increase the likelihood of benefit and result in an "acoustically transparent" fitting, conventional CROS should be configured such that sound transmitted from the poorer ear/transmitter side matches the REUG or REUR of the better ear.
8. Although CROS can help overcome a number of difficulties associated with UHL by increasing awareness of sound originating on the poorer ear side, improvement in perceptual areas requiring binaural cues (e.g., localization) is not expected.

Suggested Reading

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58 Hearing Instrument Troubleshooting Using Real Ear Measurements

John Pumford

AL is a 68-year-old female recently fitted bilaterally with open-fit behind-the-ear (BTE) hearing instruments who reports an overall lack of clarity and minimal benefit when using the devices in various listening environments.

58.1 Clinical History and Description

AL, a 68-year-old female, was recently referred by a local physician for a second opinion on the performance of her hearing instruments. AL reports recently moving to the area and indicated she is having difficulty with her recently purchased open-fit BTE hearing instruments obtained at a hearing aid center in her former community. AL indicates she has not found significant improvement in her hearing following purchase of the current devices. Overall, AL reports a lack of clarity (“muffled, unclear, dull”), continued difficulty recognizing what people are saying, and the presence of feedback. By the time of her visit, AL reports she infrequently uses her devices due to the lack of benefit. AL provided a copy of her previous audiological test results. AL reports no perceived change in her hearing sensitivity over the past several years and notes no change in her health status since she acquired her hearing instruments. A repeat audiological examination is completed to provide a current assessment of AL’s hearing status. In addition, electroacoustic analysis of the hearing instruments is completed to determine whether her devices are functioning appropriately.

58.2 Audiological Testing

Otoscopic examination was unremarkable bilaterally. Pure-tone air and bone conduction audiometry indicated a mild sloping to moderately severe sensorineural hearing loss bilaterally (Fig. 58.1). Tympanometry revealed normal tympanic membrane performance bilaterally. Speech recognition thresholds (SRTs) were consistent with pure-tone results and revealed a mild loss in the ability to receive speech. Word recognition scores (WRSs), conducted at a most comfortable listening level, revealed slight difficulty in the ability to recognize speech bilaterally. The results from the current audiological examination are consistent with those provided by AL. Electroacoustic evaluation and listening checks were performed on her hearing instruments to determine whether the devices are performing appropriately. In addition, real ear measurements (REMs) were performed to assess the audibility provided by AL’s left (Fig. 58.2) and right (not shown) devices for a range of input levels to determine whether any modification to AL’s hearing instrument settings are required.

58.3 Questions to the Reader

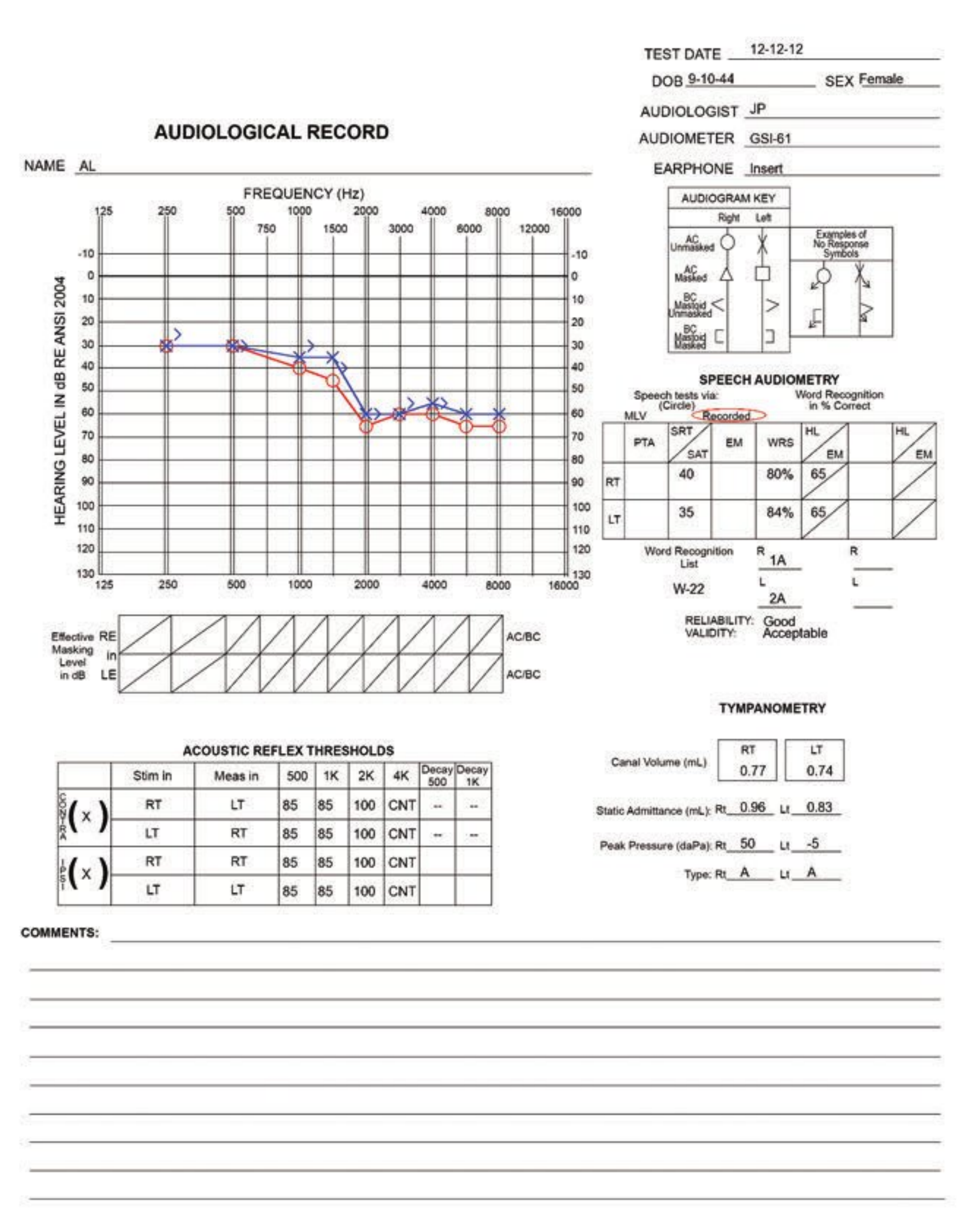
1. Which procedures and tests are most appropriate to troubleshoot AL’s concerns regarding the performance of her hearing instruments?
2. What are some considerations when conducting REMs that will improve the accuracy of the measurements?
3. Based on AL’s current hearing instrument settings, which frequency regions would be predicted to be audible/inaudible for soft (55 dB sound pressure level [SPL]), average (65 dB SPL), and loud (75 dB SPL) speech?
4. Beyond further adjustment of settings, what else could be modified with the hearing instruments to address AL’s concerns about feedback and still provide improved audibility of low-frequency sounds?

58.4 Discussion of Questions to the Reader

1. Which procedures and tests are most appropriate to troubleshoot AL’s concerns regarding the performance of her hearing instruments?

   Given that this is the first opportunity to assess AL’s hearing instruments, it is important as a first step to conduct an initial quality control assessment of her devices. This process includes a visual inspection of the hearing instruments for any noticeable damage, dirt, or debris that could impact device function (e.g., microphone screens, wax protection system, moisture in tubing, corrosion in battery compartment) along with any cleaning of components (e.g., sound bore). In addition, a listening check may identify any unusual sound quality issues/intermittencies. The listening check includes the following steps:
   a) Test the hearing instrument battery using a battery tester and replace as necessary.
   b) Attach the hearing instrument to a stethoscope and turn it on.
   c) Hold the hearing instrument a short distance (e.g., 1–1.5 ft) from the mouth and speak in a normal voice into the device while changing the user controls (e.g., volume control).
   d) Listen to the hearing instrument as you say the Ling-6 sounds (i.e., /m/, /ah/, /ee/, /oo/, /sh/, /s/).
   e) Ensure the hearing instrument sounds clear and is free of static, distortion, crackling, intermittency, or internal feedback.

   An electroacoustic analysis of the hearing instruments (e.g., ANSI 3.22 2003) would be helpful as a quality-control check and would serve as a future benchmark to ensure the devices are performing to manufacturer specifications. To ensure an accurate comparison to manufacturer specifications, the hearing instruments must be measured in the same fashion as the manufacturer (e.g., 2 cc coupler, ANSI test mode settings). The electroacoustic analysis of the hearing instruments could also be supplemented with specific tests designed to evaluate individual device components or algorithmic features (e.g., telecoil, directional microphones, noise reduction). Readers interested in



AUDIOLOGICAL RECORD

NAME AL

TEST DATE 12-12-12

DOB 9-10-44 SEX Female

AUDIOLOGIST JP

AUDIOMETER GSI-61

EARPHONE Insert

FREQUENCY (Hz)
125
250
500
750
1000
1500
2000
3000
4000
6000
8000
12000
16000
HEARING LEVEL IN dB RE ANSI 2004
-10
0
10
20
30
40
50
60
70
80
90
100
110
120
130
AUDIOGRAM KEY
Right
Left
AC Unmasked
AC Masked
BC Mastoid Unmasked
BC Mastoid Masked
Examples of No Response Symbols
Effective Masking Level in dB
RE
in
LE
AC/BC
AC/BC

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		40		80%	65		
LT		35		84%	65		

Word Recognition List W-22

R 1A R

L 2A L

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	0.77	0.74

Static Admittance (mL): Rt 0.96 Lt 0.83

Peak Pressure (daPa): Rt 50 Lt -5

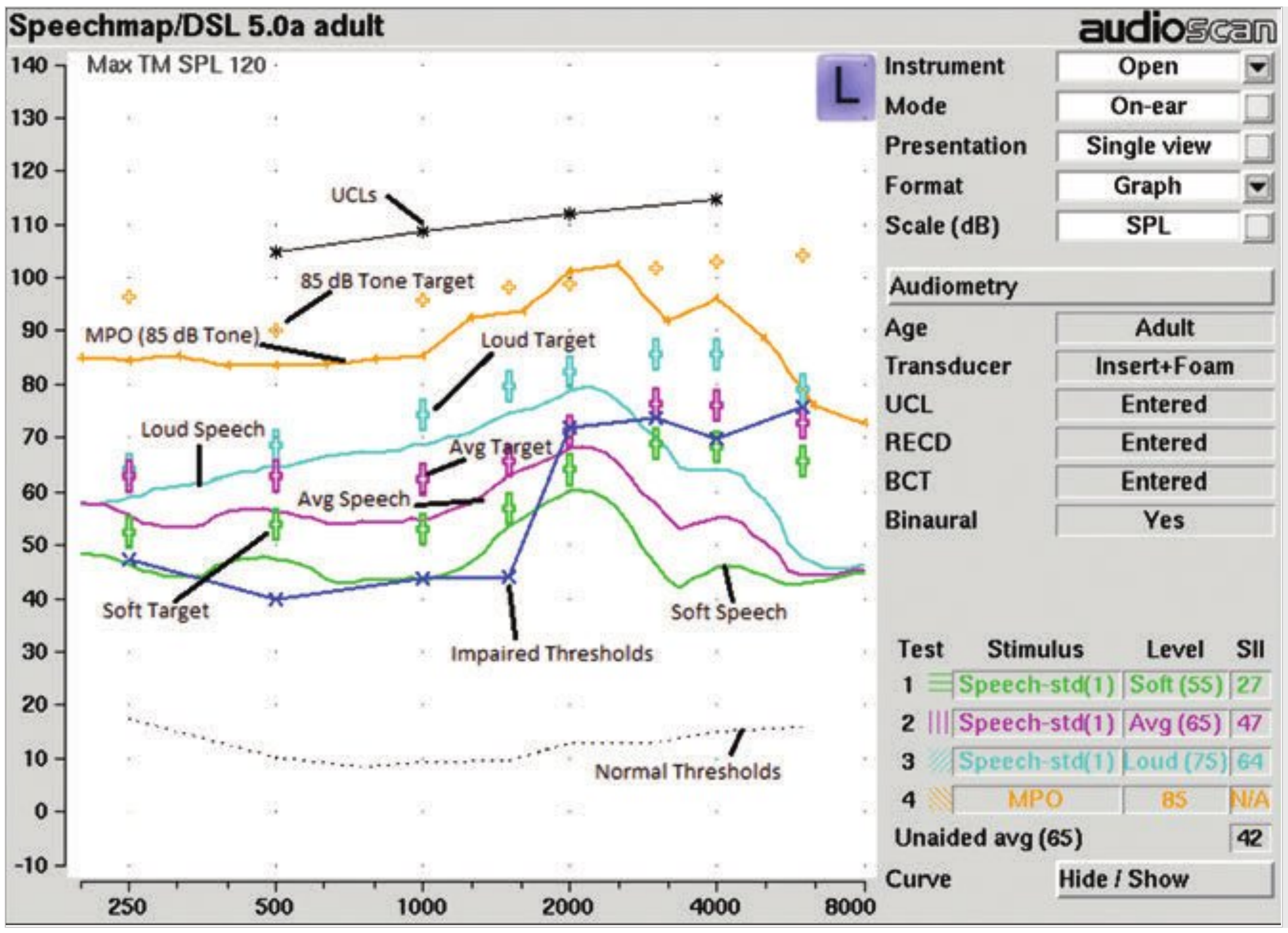
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (X)	RT	LT	85	85	100	CNT	--	--
	LT	RT	85	85	100	CNT	--	--
IPSI (X)	RT	RT	85	85	100	CNT		
	LT	LT	85	85	100	CNT		

COMMENTS:

Fig. 58.1 AL's audiological examination results.



Speechmap/DSL 5.0a adult

Instrument: Open

Mode: On-ear

Presentation: Single view

Format: Graph

Scale (dB): SPL

Audiometry

Age: Adult

Transducer: Insert+Foam

UCL: Entered

RECD: Entered

BCT: Entered

Binaural: Yes

Test	Stimulus	Level	SII
1	Speech-std(1)	Soft (55)	27
2	Speech-std(1)	Avg (65)	47
3	Speech-std(1)	Loud (75)	64
4	MPO	85	N/A

Unaided avg (65) 42

Curve: Hide / Show

Fig. 58.2 Initial real ear measurements obtained with AL's left hearing instrument, plotted in dB sound pressure level (SPL) (ear canal level) as a function of frequency. Shown in the SPL-o-Gram are normal thresholds (lower dotted line) along with AL's thresholds ("X") and uncomfortable loudness levels (UCLs) (*) for the left ear. Also displayed are real ear aided responses (REARs) for soft, average, and loud speech, the corresponding REAR targets (large "+") for these input levels, the real-ear saturation response (RESR) and corresponding RESR targets (small "+") for an 85 dB SPL pure-tone input.

learning more about these specialized tests could refer to Dillon. Most importantly, REMs would enable the clinician to determine the appropriateness of the hearing instrument settings relative to AL's hearing loss and whether her lack of benefit can be attributed to an inadequate fitting.

2. What are some considerations when conducting REMs that will improve the accuracy of the measurements?
   As with any test procedure, conclusions are only as accurate as the measurements upon which they are based. To increase the likelihood of accurate REMs, there are a number of factors that should be kept in mind, including the following:
   a) Probe tube calibration: This process mathematically accounts for the acoustic effects introduced by the probe tube and microphone during REM. Thus any error in calibration would introduce subsequent error in the displayed measurements.
   b) Probe tube placement: Ideally the end of the probe tube should be within ~ 5 mm of the tympanic membrane to minimize the impact of standing waves in the ear canal and ensure accurate measurement of high-frequency energy reaching the tympanic membrane. In this regard, otoscopy is very important to ensure proper placement and document any conditions (e.g., cerumen) that could impact the accuracy of the results.
   c) Loudspeaker location: Although typically located at a 0.5 to 1 m distance from the patient at 0° azimuth, the clinician should follow the manufacturer's recommendations regarding placement of the loudspeaker to avoid negatively impacting results.
   d) Test environment: Ideally one would select a relatively quiet environment to minimize the influence of environmental noise on measurements, particularly for softer input signals. In addition, placement of the test equipment and client away from any reflective surfaces would minimize the likelihood of standing waves or sound reflections adversely influencing the measurement. To properly account for these environmental concerns, guidelines outlined by Revit state that the distance from both the subject and the loudspeaker to any reflective surface should be at least twice the distance between the subject and the loudspeaker (i.e., twice the "working distance").
   e) Signal type: Ideally a speech or speechlike test signal having the appropriate frequency, intensity, and temporal characteristics of speech would be used. Should only non-speech verification signals (e.g., noise, pure-tones) be available, the clinician should disable adaptive features (e.g., noise reduction, phase canceler) or select the hearing aid manufacturer's verification mode to avoid creating atypical measurement artifacts.
   f) Sound calibration method: To ensure the input level received at the hearing aid microphone location is correct, REM systems may use an external reference microphone to monitor the sound field at the measurement point (i.e., ear level) and adjust the loudspeaker level accordingly. With open-fit products, amplified hearing aid sound escaping the ear canal via the vent can be picked up by this reference microphone and result in an adjustment of the loudspeaker level. To avoid contamination of REMs, clinicians should use stored equalization rather than concurrent equalization during testing with open-fit products. This process disables the reference microphone during the measurement, but not the leveling procedure, and ensures sound leaking from the ear canal will not affect the loudspeaker level and the measurement. A full discussion of REM factors is beyond the scope of this case report, but interested readers could refer to Mueller et al, Revit, Pumford and Sinclair, or Mueller and Ricketts for more details.
3. Based on AL's current hearing instrument settings, which frequency regions would be predicted to be audible/inaudible for soft (55 dB SPL), average (65 dB SPL), and loud (75 dB SPL) speech?
   REMs conducted with AL's hearing instruments at their initial settings indicated that the devices were not providing sufficient audibility. Results indicated underamplification relative to Desired Sensation Level (DSL) v.5 adult targets ("+" symbols) for all input levels for the left (□ Fig. 58.2) and right (not shown) hearing instruments. The real ear aided responses (REARs) for the left ear (□ Fig. 58.2) indicated that audibility was provided from ~ 250 to 1500 Hz for soft (green curve) speech, to ~ 2 Hz for average (pink curve) speech and up to ~ 3000 Hz for loud (blue curve) speech. Measured REARs for the right ear were similarly problematic, indicating little to no audibility for soft speech from 250 to 8000 Hz, audibility to ~ 1500 Hz for average speech, and audibility to ~ 2000 Hz for loud speech. This finding of insufficient high-frequency output across input levels was consistent with AL's audibility and clarity complaints. Although the basis for AL's current hearing instrument settings were unclear, past research has shown that an over-reliance on first-fit/quick-fit programming software logic can result in measured output levels that are significantly different than fitting formula targets.

   The real ear saturation responses (RESRs; orange line) for the left (□ Fig. 58.2) and right (not shown) hearing instruments did not exceed the associated 85 dB tone targets (orange +) or AL's measured uncomfortable loudness levels (UCLs) (black *). Although it is important to ensure that hearing aid output does not exceed an output level that will cause loudness discomfort, it is also equally important to ensure the maximum output of the device is not overly restrictive because it can negatively impact speech recognition and sound quality. As shown in □ Fig. 58.2, the RESR for the left device fell significantly below the associated 85 dB tone targets from ~ 250 to 2000 Hz and from ~ 3000 to 8000 Hz. Similar results were obtained with the right device (not shown). With these considerations in mind, the settings of the devices were adjusted to provide a more appropriate output for AL's hearing loss. This process initially resulted in a closer match to DSL v.5 adult targets bilaterally. Product performance, however, was still deemed to be unsatisfactory. First, AL began to experience occasional feedback at the higher output settings. Second, despite a significant increase in the low-frequency output settings in the left hearing instrument, low-frequency REAR targets still could not be achieved. In an attempt to address the feedback concern, a static feedback test was performed using the manufacturer's fitting software. As reported in □ Fig. 58.3, given the open-vent condition, the maximum stable gain in the fitting before feedback was determined to be significantly lower than the gain available in each device. Further, the application of this gain limit

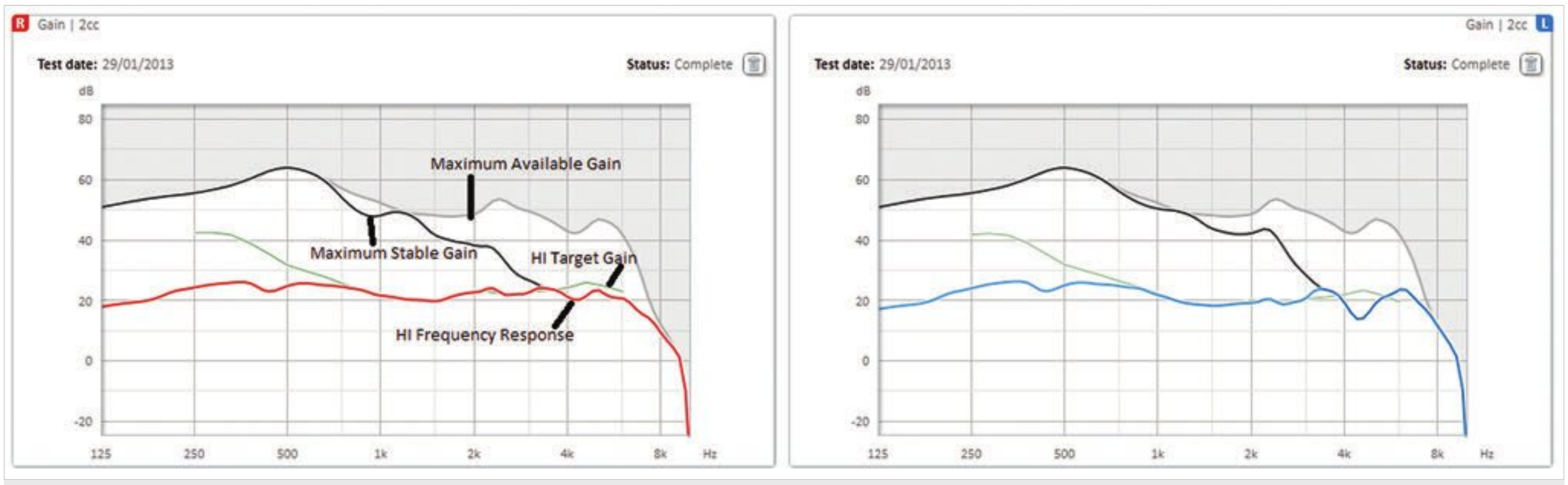


Fig. 58.3 Feedback optimization measurements conducted with AL's left and right hearing instruments with open dome coupling. Shown are the maximum available gain, the maximum stable gain, target gain, and the frequency response.

in the software resulted in a high-frequency output reduction in the hearing instruments, an inability to adjust settings higher than this gain limit, and ultimately an underfitting of the desired high-frequency REAR targets.

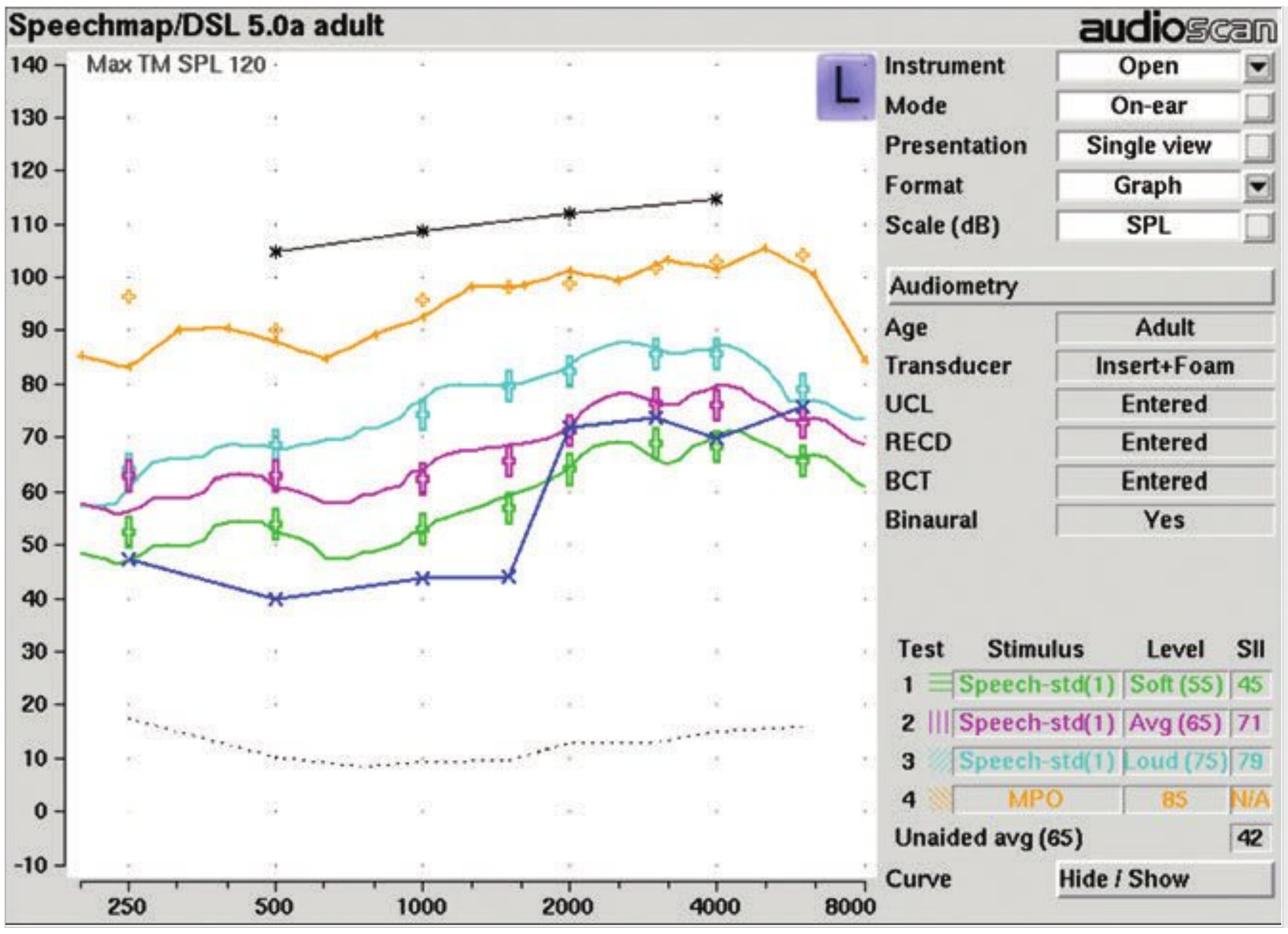
4. Beyond further adjustment of settings, what else could be modified with the hearing instruments to address AL's concerns about feedback and still provide improved audibility of low-frequency sounds?
   Adjustment of the venting/earpiece coupling attached to AL's hearing instrument would be one method to address feedback concerns and low-frequency audibility requirements. Specifically, consideration of the current coupling provided for AL suggested a need to provide a more occluding dome. This change would result in less sound being vented from the hearing instruments, thereby providing a potentially positive impact on maximum stable gain (i.e., decreased likelihood of feedback) and the ability to provide additional low-frequency amplification. The potential benefits of providing a more occluding earpiece in this case, however, needed to be balanced with the potential negative consequences of creating occlusion issues given AL's normal/near normal low frequency hearing sensitivity. Dillon reported that vent size should be selected such that the prescribed target output can be achieved without introducing feedback or excessively occluding the ear canal. With these factors in mind, the coupling was modified from an "open" dome to a slightly more occluding "closed" dome, and the vent parameter in the hearing instrument manufacturer software was adjusted accordingly to ensure the modified venting characteristics were accounted for in the gain/frequency response calculation. The static feedback test performed following this change was consistent with a more occluding vent condition because the maximum stable gain limit in the high-frequency region was significantly greater than that observed in Fig. 58.3 for both devices.
   To assess whether the reduced venting would create issues with occlusion, an occlusion effect test was performed bilaterally using the Verifit (Audioscan, Dorchester, ON, Canada). First, a probe tube is placed in the ear canal and the client is asked to vocalize the sound /ee/ (i.e., back vowel generating low-frequency energy in the ear canal) without a hearing instrument in place (i.e., unaided SPL in ear canal is measured). Second, the hearing instrument is inserted into the ear canal (while turned off), and the REM is repeated as the client vocalizes /ee/ at the same level generated previously. A comparison is made between the REM obtained while the ear canal is unoccluded versus occluded, with any difference reflecting the magnitude of the occlusion effect. Occlusion effect testing did not suggest any concerns with occlusion bilaterally as the two measurement curves were superimposed. AL's comments regarding her voice with the hearing instruments in place also did not suggest any concerns with occlusion. Subsequent adjustments to the hearing instruments settings were performed to provide improved audibility and a closer match to the prescribed DSL v.5 adult REAR and RESR targets for the left (Fig. 58.4) and right (not shown) devices.

58.5 Diagnosis and Recommended Treatment

A visual inspection of AL's hearing instruments did not reveal any concerns with dirt or debris, and an initial listening check did not reveal any artifacts or intermittencies. An electroacoustic evaluation indicated both devices were functioning appropriately when compared with the manufacturer's ANSI specifications. Based on the aforementioned REMs (Fig. 58.2) it was determined that both hearing instruments were not providing sufficient audibility in the frequency region above ~2000 Hz for soft, average, and loud speech input levels at the initial settings. Subsequent adjustment of the output/frequency response of AL's hearing instruments in combination with a change to a slightly more occluding closed dome resulted in improved audibility and a closer match to DSL v.5 adult REAR and RESR targets for the left (Fig. 58.4) and right (not shown) devices. AL subsequently reported improved audibility of speech in the clinic versus her initial settings and acceptable sound quality for her own voice.

58.6 Outcome

A follow-up call with AL 1 week following her appointment indicated she was performing well with her hearing instruments following the aforementioned adjustments. AL reported



Instrument: Open
Mode: On-ear
Presentation: Single view
Format: Graph
Scale (dB): SPL

Age: Adult
Transducer: Insert+Foam
UCL: Entered
RECD: Entered
BCT: Entered
Binaural: Yes

Test	Stimulus	Level	SII
1	Speech-std(1)	Soft (55)	45
2	Speech-std(1)	Avg (65)	71
3	Speech-std(1)	Loud (75)	79
4	MPO	85	N/A
Unaided avg (65)			42

Fig. 58.4 Real-ear measurements obtained with AL's left hearing instrument with "closed" dome coupling following completion of the feedback test and subsequent fine tuning, plotted in dB SPL (ear canal level) as a function of frequency. Shown are normal thresholds (lower dotted line) along with AL's thresholds ("X") and uncomfortable loudness levels (UCLs) (*) for the left ear. Also displayed are real ear aided responses (REARs) for soft, average, and loud speech, the corresponding REAR targets (large "+") for these input levels, the real-ear saturation response (RESR), and corresponding RESR targets (small "+") for an 85 dB SPL pure-tone input.

she was hearing significantly better with her hearing instruments and perceived a noticeable improvement in clarity, sound quality, and speech recognition in various listening environments. No significant concerns with her own voice were expressed, and AL indicated no further concerns with her hearing instrument performance.

58.7 Key Points

1. REM is an invaluable troubleshooting tool that allows a clinician to quickly, systematically, and accurately determine the appropriateness of a patient's hearing instrument fitting relative to his or her residual dynamic range. Using this information, the audiologist can determine the appropriate course of action to resolve any identified issues based on objective evidence.
2. To increase the likelihood of accurate REMs, the impact of various factors should be kept in mind, including probe-tube calibration, probe-tube placement, loudspeaker location, test environment, signal type, and method of sound calibration.
3. Hearing instrument setting changes alone may not be sufficient to address listening issues. The impact of nonelectroacoustic factors on hearing instrument performance should not be overlooked. For instance, factors such as venting/earpiece coupling should be considered and modified systematically when trying to find the proper balance between audibility, feedback resolution, and avoidance of occlusion.

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59 Real Ear Measures Were Completed, but. . .

Michael Valente and L Maureen Valente

Two national guidelines are available supporting the use of real ear measures (REMs) to verify the performance of hearing aids. Unfortunately, approximately 70 to 75%of audiologists still do not routinely verify hearing aid performance using REMs, but instead typically download the manufacturer's First-Fit to the hearing aids. This is despite an abundance of evidence reporting that the First-Fit usually underamplifies the high-frequency region of the hearing aid's frequency-gain and output response. Even for those relatively few audiologists who do complete REMs, this case report highlights some "finer nuances" of REMs that some students and clinicians may not be aware of.

59.1 Clinical History and Description

JR is a 52-year-old male who presented to the audiology clinic with a primary interest in pursuing amplification. He reports long-standing bilateral hearing loss. JR reports of greater difficulty recognizing female and child talkers, recognizing the speech of female news reporters while watching television, recognizing speech in noisy listening situations, and communicating on the telephone. He denies tinnitus, dizziness, vertigo, history of exposure to noise, or previous otologic surgery.

59.2 Audiological Testing

The results from the audiological examination reveal a bilateral symmetrical moderate to severe sensorineural hearing loss (SNHL) at 250 to 8000 Hz that is gradually sloping in configuration. The bilateral speech recognition thresholds (SRTs) of 60 dB HL reveal a moderate loss in the ability to receive speech and are in agreement with the pure-tone averages (PTAs). The word recognition scores (WRSs) at JR's most intelligible level (MIL) using the full-list female version of the recorded Northwestern University Auditory Test No. 6 (NU-6) word lists were 84% bilaterally. These results reveal JR's slight difficulty in the ability to recognize speech bilaterally. Immittance audiometry reveals normal bilateral tympanograms. Ear canal volumes were 0.7 and 0.8 mL, static compliance measures were 0.6 and 0.8 mL, and middle ear pressure was 0 and − 10 daPa for the right and left ears, respectively. These finding indicate normal tympanic membrane and middle ear function. Bilateral acoustic reflex thresholds to contralateral and ipsilateral stimulation at 500 to 4000 Hz and reflex decay at 500 and 1000 Hz are indicative of cochlear involvement. A cover letter, summary report, and copy of the audiometric examination were faxed to JR's primary care physician recommending that JR return for a hearing aid evaluation (HAE) following a referral to an otologist for medical clearance for amplification. JR was seen by his otologist who provided medical clearance, and JR was scheduled for an HAE.

59.2.1 Hearing Aid Evaluation

Following the audiological examination and medical clearance an HAE was completed. During the HAE the following items were reviewed and discussed:

1. The audiologist reviewed JR's audiological results because several weeks had passed since the initial audiological examination. Experience has shown that most patients forget the counseling that was completed at the audiological examination and a review is often very helpful.
2. Prior to the appointment, the audiologist calculated JR's Speech Intelligibility Index (SII) using a free web-based program (http://sii.to/). In JR's case the calculated SII was 0.02 using the 1/3 octave method, which indicates that only 2% of average conversational speech is available to JR. Reporting and explaining JR's calculated SII allowed JR to have a greater appreciation for the impact his hearing loss has on his increased communication difficulty. Areas of difficulty include recognizing speech (1) of female and child talkers, (2) of female news reporters on television, (3) in noisy listening situations, and (4) on the telephone.
3. The audiologist reviewed the anatomy and physiology of the ear using a variety of figures that have been accumulated from textbooks and journals. This review allowed JR to have a greater appreciation of how his hearing loss is related to anatomical changes in his inner ear; his outer and middle ear is normal.
4. The audiologist counseled JR on the advantages of bilateral amplification for improved hearing in background noise, localization, and spatial balance.
5. A clinic-created counseling tool using a bar graph was used to illustrate realistic expectation from amplification. This bar graph plots unaided and aided "problem scores" (the higher the bar, the poorer the performance) for speech recognition in quiet, noise, and reverberation. In this graph, the height of the bar for the unaided problem score for each of the three listening conditions is significantly higher than the height of the aided score, but no aided problem score approaches normal (i.e., 0%problem score). This illustrates to JR that aided performance is better than unaided performance, but aided performance will not be the same as when JR had normal hearing. Further, JR's aided performance in noise and reverberation is better (lower bar) than his unaided performance (higher bar), but his aided performance for these two difficult listening conditions will always be poorer (bar is higher) than the aided performance for the quiet listening condition. That is, aided performance in noise will be better than unaided performance in noise, but aided performance in noise will never be as good as JR's aided performance in quiet. It was emphasized that even listeners with normal hearing have poorer performance in noise than in quiet.
6. The audiologist demonstrated different hearing aid styles to JR. This included custom hearing aids, conventional

behind-the-ear (BTE) instruments, and BTEs with receivers in the hearing aid using a thin-tube to direct the amplified sound to the ear canal or the use of an ear wire connected to the receiver placed in the ear canal. JR was also shown examples of manufacturer domes and custom earmolds that can be used to deliver the amplified sound to the ear canal. It was emphasized to JR that custom earmolds are more expensive than domes (which are free), but custom earmolds may provide greater comfort and retention and allow for possibly greater amplification before feedback becomes a problem.

7. The audiologist discussed different features of hearing aids such as volume controls, program buttons, remote control, and directional microphones. The audiologist also discussed wireless communication to cellular and landline telephones, TV, hearing assistive technology, and a wide variety of other auditory inputs.
8. The audiologist discussed and explained the importance of the telecoil for communication on the telephone and hearing assistive technology.
9. JR was counseled on the relationship among the levels and associated costs of technology. These differences in levels include the number of available bands (flexibility in the ability for the clinician to match a validated prescriptive target using REMs) and channels (effectiveness of the signal processing in processing environmental noise and feedback as well as the effectiveness of the directional microphones), automatic features, warranty for repairs (1–3 years), and loss and damage (1–3 years).
10. JR was provided with the clinic hearing aid information sheet that outlines how hearing aids are dispensed in this clinic.
11. The audiologist counseled JR on the clinic's policy for providing hearing aid maintenance checks and future reprogramming of the hearing aids.
12. The audiologist administered the unaided Client-Oriented Scale of Improvement (COSI) with JR. For his responses, JR wrote that he wanted his hearing aids to improve his ability to recognize (1) female and child talkers, (1) female news reporters on television, (3) speech in noisy listening situations, and (4) communication on the telephone.
13. The audiologist measured loudness discomfort levels (LDLs) bilaterally in dB HL and sound pressure level (SPL) with a probe tube placed in the ear canal at 500 to 4000 Hz. The measured LDL was ascertained when JR stated the test signal was "loud, but OK." These LDL measures served as the target when measuring the real ear saturation response to a pure-tone sweep presented at 90 dB SPL (RESR90) during the subsequent hearing aid fitting (HAF).
14. Upon completing the HAE, bilateral hearing aids with 20 channels of signal processing were recommended and subsequently ordered. Earmold impressions were completed to order bilateral custom earmolds with select-a-vent (SAV).
15. Finally, the clinic received and scanned JR's medical clearance for amplification into JR's electronic medical record (EMR).

59.2.2 Hearing Aid Fit

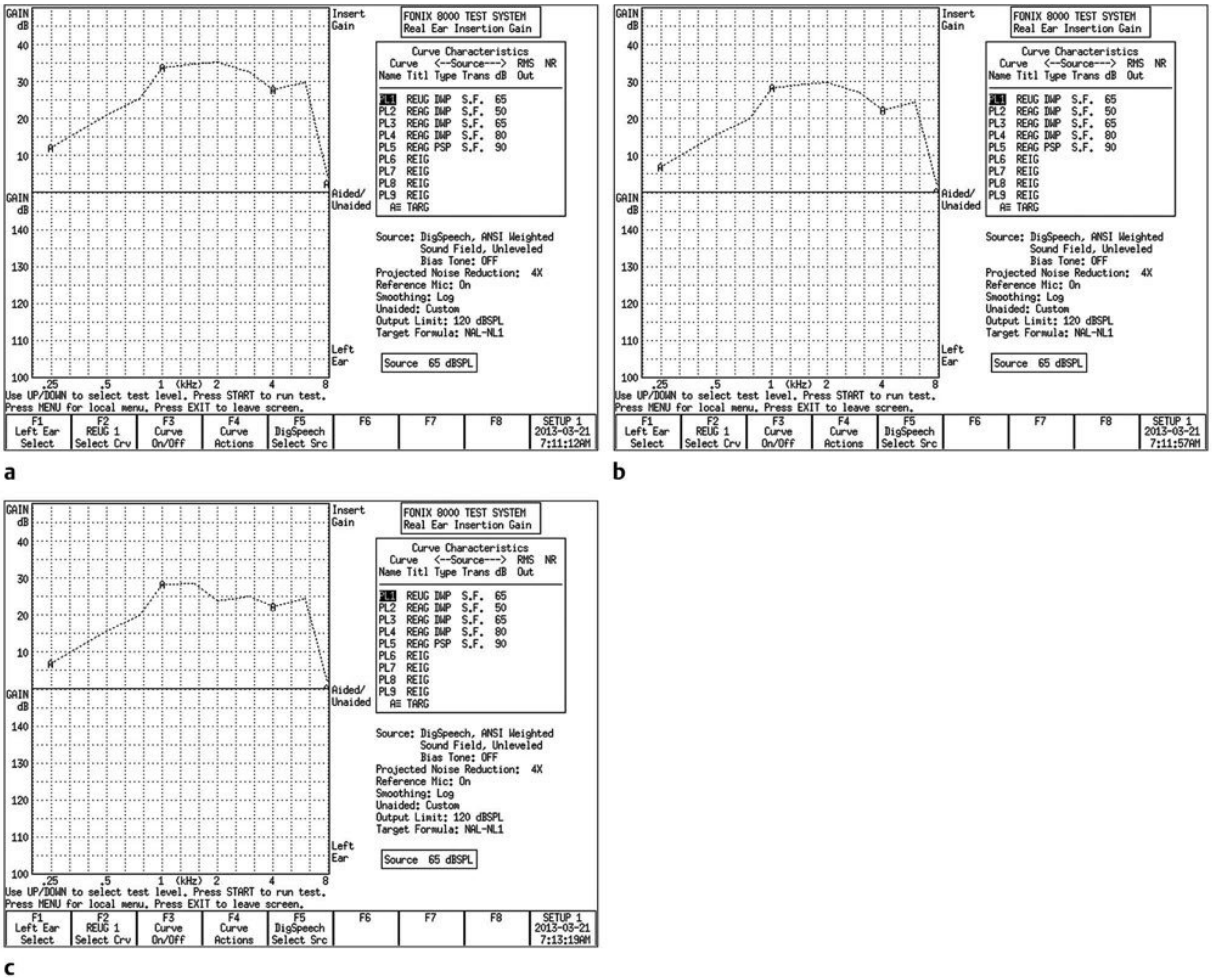
JR was fit with bilateral hearing aids with 20 channels of signal processing and custom earmolds that were appropriate for his hearing loss. As an initial step for verifying the fitting, JR's bilateral audiometric thresholds were entered into the software of the Frye 8000 real ear analyzer (Frye Electronics, Inc., Tigard, OR). In the Frye 8000 system, the default setting assumes a unilateral fit for a hearing aid with one channel of signal processing. Fig. 59.1a reports the prescribed real ear insertion gain (REIG) for the left ear using National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) as the prescriptive target. As the reader recalls only 25 to 30% of audiologists arrive at this point, and some of those would use the REIG in the upper curve of Fig. 59.1 to serve as the prescribed target. The audiologist proceeds to manipulate the hearing aid manufacturer's software to program the hearing aids so the measured REIG arrives as close to this target as possible. Fig. 59.1b, however, reports the prescribed REIG after correcting for a bilateral fit within the software of the Frye 8000. In examining the upper and middle graphs of Fig. 59.1 it can be seen that at all frequencies the corrected REIG for the bilateral fit reduces the prescribed REIG by 5 dB at each frequency. Next, the audiologist reentered the menu on the Frye 8000 system and changed the number of channels from one channel to 18+ channels (in the 8000, the maximum number of channels is 18+). Fig. 59.1c reports the prescribed REIG corrected for bilateral and channel summation.

In comparing the lower graph of Fig. 59.1 to the upper graph of Fig. 59.1 it can be seen that the difference between the "final" corrected REIG (Fig. 59.1c) and the "original" REIG (Fig. 59.1a) ranges from 5 to 11 dB depending upon frequency. If the audiologist were not aware of the need to correct the prescribed REIG for the bilateral fit and the 20 channels of signal processing and instead programmed the hearing aids to the original prescribed REIG JR would report that his hearing aids are "too loud."

Going one step further, as stated earlier, approximately 70 to 75% of audiologists do not routinely use REMs to verify the performance of hearing aids. Those audiologists typically enter the patient's hearing thresholds into NOAH. These thresholds are incorporated into the manufacturer's proprietary fitting formula to create the manufacturer's recommended First-Fit. In this case, these results would be downloaded to each of JR's hearing aids. At this point, the audiologist would typically ask the patient how the hearing aid sounds, and that would be the extent of verification. This is often the norm in spite of numerous articles in peer-reviewed journals that have repeatedly reported that the gain/output provided by the typical manufacturer First-Fit results in a frequency-gain response that significantly underamplifies the prescribed target in the higher frequencies. To emphasize this point in this case report, Fig. 59.2a reports the manufacturer First-Fit using the manufacturer's version of the NAL-NL1 target to program the hearing aids. As can be seen, the manufacturer's First-Fit significantly underamplifies the target at 1500 Hz and above. Fig. 59.2b, on the other hand, reports the final programmed fit to the NAL-NL1 target using 65 dB SPL modulated DigiSpeech speech-weighted noise. Abrams et al reported significantly better outcomes when hearing aid users were fit to a prescribed target than when the hearing aids were fit using First-Fit.

59.3 Questions to the Reader

1. Does every real ear system default to a unilateral fit and a hearing aid with single-channel signal processing?



a

```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1  REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3  REAG DWP  S.F.  65
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7  REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Unleveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-03-21
7:11:12AM
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b

```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1  REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3  REAG DWP  S.F.  65
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7  REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Unleveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-03-21
7:11:57AM
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c

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FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1≡ REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3  REAG DWP  S.F.  65
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7  REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Unleveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-03-21
7:13:19AM
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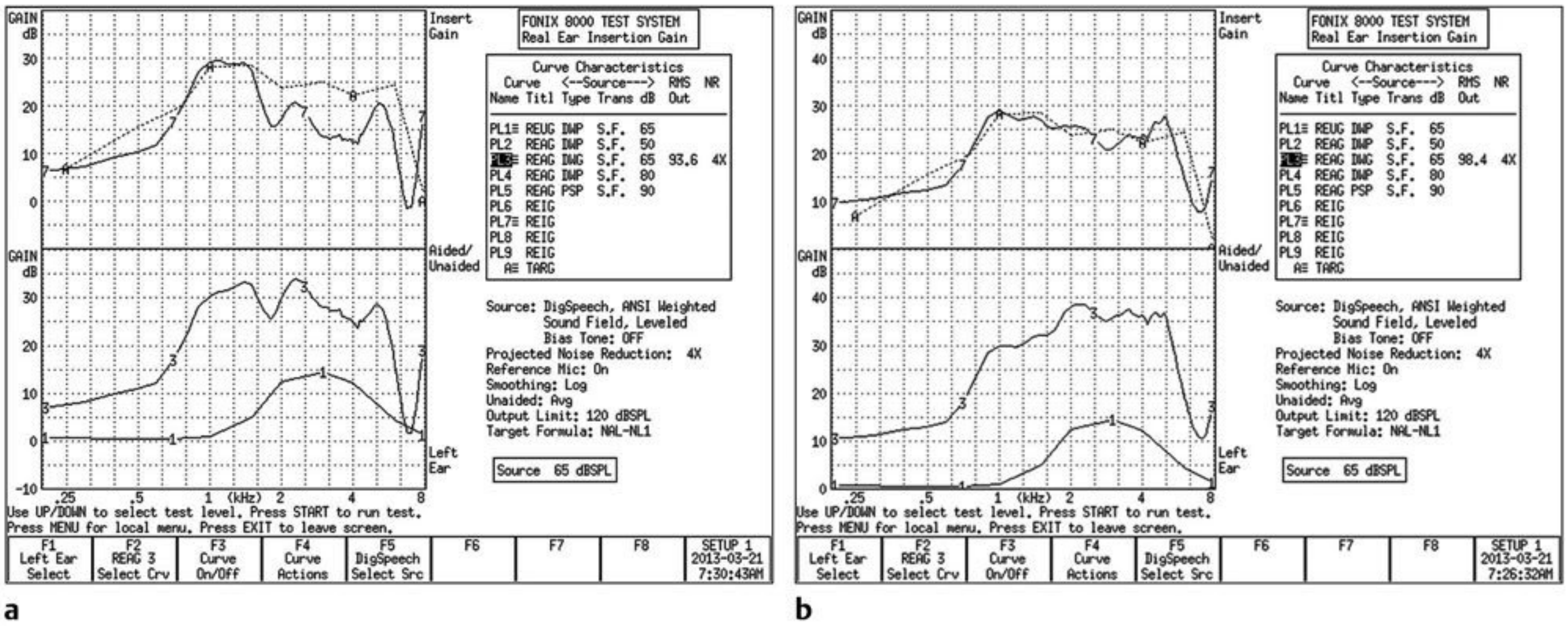
Fig. 59.1 (a) The prescribed real ear insertion gain (REIG) target for the left ear using National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) as the prescriptive target. (b) The prescribed REIG after correcting for a bilateral fit. (c) The prescribed REIG after correcting for binaural and channel summation.



a

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FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1≡ REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3≡ REAG DWG  S.F.  65  93.6  4X
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7≡ REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Avg
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-03-21
7:30:43AM
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b

```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out
PL1≡ REUG DWP  S.F.  65
PL2  REAG DWP  S.F.  50
PL3≡ REAG DWG  S.F.  65  98.4  4X
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7≡ REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Avg
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-03-21
7:26:32AM
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Fig. 59.2 (a) The manufacturer First-Fit to the corrected real ear insertion gain (REIG) target. (b) The programed fit to the corrected REIG target using 65 dB sound pressure level of modulated DigiSpeech speech-weighted noise.

2. What is the purpose of showing JR a bar graph illustrating unaided and aided performance with hearing aids in the three listening conditions?
3. How would a clinician know how many channels of signal processing are available in the hearing aids being dispensed?
4. What is the timeframe for a current audiogram to be completed before an audiologist would be comfortable going forward with fitting hearing aids.
5. When clinicians use REMs to verify hearing aid fittings using real ear aided response (REAR) measures instead of REIG, do the corrections for bilateral fitting and the number of channels also apply to REAR measures?

59.4 Discussion of Questions to the Reader

1. Does every real ear system default to a unilateral fit and a hearing aid with single-channel signal processing?
   The authors have much of their clinical experience in REMs using Frye equipment and are not as knowledgeable with the software accompanying the other REM systems that are commercially available. The student or clinician should consult the manual for the REM system being used or contact the manufacturer to provide an answer to this question.
2. What is the purpose of showing JR a bar graph illustrating unaided and aided performance with hearing aids in the three listening conditions?
   In the experience of the authors, one of the primary causes for rejection of amplification, especially for the first-time user, is unrealistic expectations of hearing aids. The counseling tool described in this case report was developed to counsel a patient regarding reasonable expectations of hearing aids. This counseling tool allows the clinician, by using "real numbers," to emphasize the point that aided performance in noise is typically better than unaided performance in noise. Aided performance in noise, however, will never be as good as aided performance in quiet. It should be emphasized that even listeners with normal hearing perform more poorly in noise than in quiet.
3. How would a clinician know how many channels of signal processing are available in the hearing aids being dispensed?
   This information can be obtained in a number of ways. First, it is available on the manufacturer specification sheet that can be obtained at the manufacturer website. Second, many of the manufacturer's NOAH software modules include product information, and the number of channels of signal processing may be viewed from within NOAH. A quick phone call to the manufacturer would provide the answer to this question.
4. What is the timeframe for a current audiogram to be completed before an audiologist would be comfortable going forward with fitting hearing aids?
   When fitting hearing aids, an accurate audiological examination must be completed no more than 6 months from the time the hearing aids are dispensed. In addition, the patient must obtain medical clearance for amplification or have signed a waiver (if an adult). At the authors' clinic, the audiologists strongly recommend a signed form for medical clearance and do not promote the signing of a waiver.
5. When clinicians use REMs to verify hearing aid fittings using real ear aided response (REAR) measures instead of REIG, do the corrections for bilateral fitting and the number of channels also apply to REAR measures?
   Yes.

59.5 Outcome

JR was very pleased with the performance of his hearing aids and custom earmolds. He reported no problems with loudness, and his aided COSI scores reported "much better" performance in his ability to recognize (1) female and child talkers, (2) female news reporters on television, and (3) speech in noisy listening situations, and "slightly better" performance for communication on the telephone.

59.6 Key Points

1. Always verify the performance of hearing aids using real ear measures. Do not rely on the manufacturer's First-Fit.
2. Always confirm the performance of hearing aids using a validated questionnaire.
3. Be aware and take the necessary steps to correct a prescriptive target for monaural versus bilaterally fittings and for the number of channels of signal processing.

Suggested Reading

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Valente M, Abrams H, Benson D et al. American Academy of Audiology guideline for the audiologic management of adult hearing impairment. Audiol Today 2006; 18: 336–343

60 Effect of Domes versus Custom Earmolds

Michael Valente, L. Maureen Valente, and Rachael Mangiore

The goal of a hearing aid fitting is to match, as closely as possible, real ear insertion gain (REIG) or the real ear aided response (REAR) to a validated prescriptive target such as National Acoustic Laboratories' nonlinear fitting formula version 1 or 2 (NAL-NL1 or NAL-NL2). With the recent introduction of hearing aids using thin-tubes or a wire connected to a receiver-in-the-canal (RIC), audiologists may couple these hearing aids to the ear canal using domes or custom earmolds. Coupling hearing aids with domes or custom earmolds may create significant differences in programming hearing aids to match the measured REIG or REAR to a validated prescriptive target.

60.1 Clinical History and Description

This case report describes a 65-year-old male with long-standing bilateral sensorineural hearing loss who inquired about amplification to improve communication with his work as an attorney. He did not report any tinnitus in either ear, recruitment, otalgia, dizziness, fullness, vertigo, excessive exposure to noise, prior experience with amplification, or otologic surgery. He reported increased difficulty recognizing speech in noise, female talkers, communicating on the telephone, and greater difficulty recognizing speech at meetings in his office.

60.2 Audiological Testing

The patient was initially seen a year earlier and returned for an annual audiological evaluation. The results from the most recent audiological evaluation revealed an asymmetric slight to moderately severe sensorineural hearing loss (SNHL) in the left ear and a slight to severe SNHL in the right ear at 500 to 8000 Hz. Hearing was within normal limits bilaterally at 250 Hz. The ears were symmetrical with the exception of a 15 dB difference at 750 and 2000 Hz, 20 dB differences at 8000 Hz, and a 25 dB difference at 1000 Hz. Relative to an audiogram obtained a year earlier, there was a 15 dB decrease in the right ear at 1000 Hz and a 10 dB decrease at 2000 Hz. In addition, the word recognition score (WRS) in the right ear decreased from 92 to 76% Hearing thresholds in the left ear remained stable, as did the WRS. Bilateral immittance, acoustic reflex thresholds to contralateral and ipsilateral stimulation at 500 to 4000 Hz, and reflex decay at 500 and 1000 Hz were within normal limits. A letter, summary report, and copy of the audiometric evaluation were faxed to the patient's primary care physician recommending a referral to an otologist for follow-up care due to the change in hearing since the previous audiometric evaluation. In addition, medical clearance for amplification was requested. The patient was referred to an otologist who ordered magnetic resonance imaging (MRI), and the results were negative. At that point, medical clearance was obtained and the hearing aid fit (HAF) was scheduled.

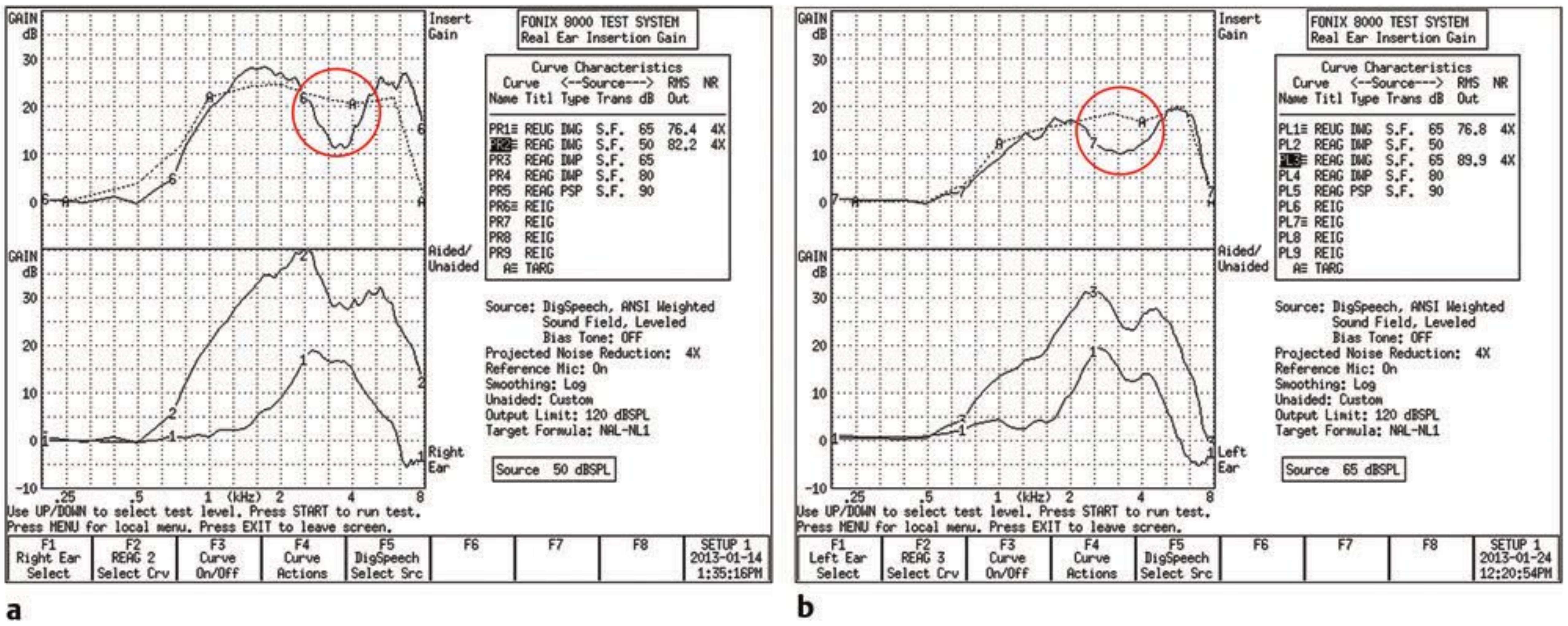
60.2.1 Hearing Aid Evaluation

Following the audiological evaluation, a hearing aid evaluation (HAE) was completed and bilateral RIC hearing aids were recommended. As part of the HAE, the patient was counseled on the manner in which the RIC hearing aids can be coupled to the ear canal. During the counseling session, the audiologists displayed domes and custom earmolds and counseled on the advantages and disadvantages of each. Because of the bias of the audiologists based on previous fittings, the advantages of custom earmolds were emphasized. Advantages discussed included greater comfort and retention as well as less probability for feedback. The patient, however, decided to initially try open domes to reduce cost, and stated he may consider ordering earmolds later if he felt the domes were uncomfortable, required constant reinsertion (i.e., retention challenges), or produced feedback.

60.2.2 Hearing Aid Fit

The patient was fit with bilateral Widex Clear 440-F RIC hearing aids (Widex USA, Inc., Hauppauge, NY) with #2 ear wires, M receivers, and an RC-DEX remote control. The RICs were coupled to the ear using open domes. Unfortunately, during real ear measures (REMs) it was seen that the measured REIG between 3000 and 4000 Hz was ~ 10 to 15 dB below the prescribed REIG for NAL-NL1 (circled areas in Fig. 60.1). In Fig. 60.1 (right and left ears) curve A represents the prescribed NAL-NL1 REIG target for a 65 dB SPL input level corrected for a bilateral fit and 15 channels of signal processing. Curve 7 is the measured REIG for an input level of 65 dB SPL using a modulated speech-weighted signal. Curves 6 and 8 represent the measured REIG for the 50 (curve 6) and 80 dB (curve 8) sound pressure level (SPL) input levels. As can be seen in Fig. 60.1, the measured REIG does not match the prescribed REIG at ~ 3000 to 4000 Hz after completing the feedback test (Fb) (Fig. 60.2a) within the Widex Compass software. As can be seen in Fig. 60.2a the amount of available gain prior to feedback in this frequency region was limited due to the software detecting the presence of feedback. Because of this, the audiologists were not able to provide any additional gain (Fig. 60.1).

At this point, the patient was counseled on the results of the Fb test and the real ear results and was reminded of the need to consider custom earmolds. This was in order to achieve the desired outcome of matching REIG to target between 3000 and 4000 Hz because this is an area very important for speech recognition. The patient agreed, bilateral silicone impressions were completed, and custom earmolds were ordered. When the earmolds arrived, the Fb was repeated (Fig. 60.2b for the right and left ears). As may be seen, the results from the Fb revealed significantly greater available gain than was allowed in the initial Fb test (Fig. 60.2a). In addition, the audiologists were able to better match the REIG target (circles in Fig. 60.3 for the right and left ears, respectively). Finally, the patient



```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
 Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out

PR1≡ REUG DWG  S.F.  65  76.4  4X
PR2≡ REAG DWG  S.F.  50  82.2  4X
PR3  REAG DWP  S.F.  65
PR4  REAG DWP  S.F.  80
PR5  REAG PSP  S.F.  90
PR6≡ REIG
PR7  REIG
PR8  REIG
PR9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

 Source  50 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
2013-01-14
1:35:16PM
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```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
 Curve   <--Source---> RMS  NR
Name Titl Type Trans dB  Out

PL1≡ REUG DWG  S.F.  65  76.8  4X
PL2  REAG DWP  S.F.  50
PL3≡ REAG DWG  S.F.  65  89.9  4X
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7≡ REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

 Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.

SETUP 1
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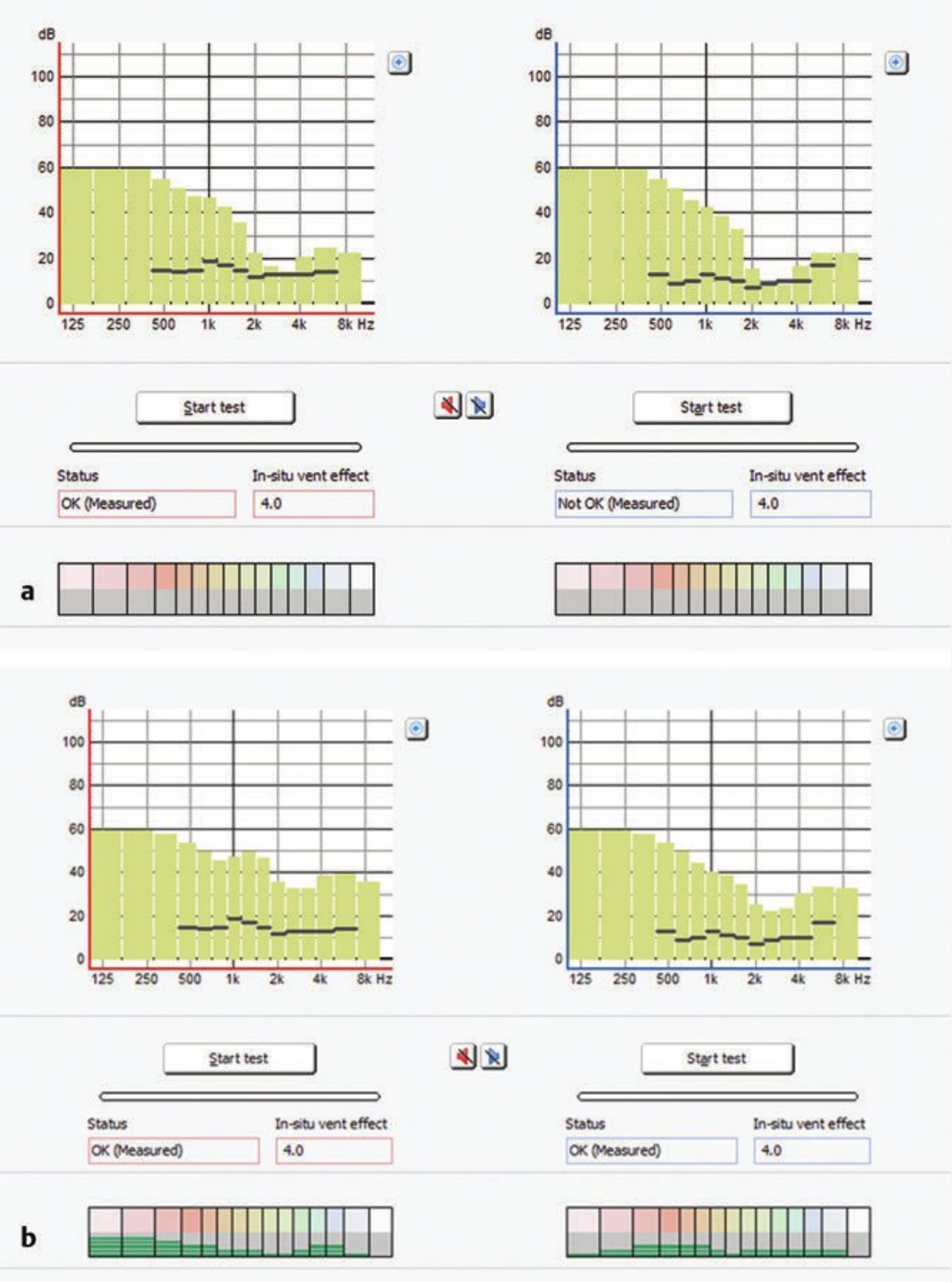
Fig. 60.1 Measured real ear insertion gain (REIG) (curve 7) and prescribed REIG (curve A) for the right (a) and left (b) ears using a dome.



a

Status: OK (Measured) — In-situ vent effect: 4.0

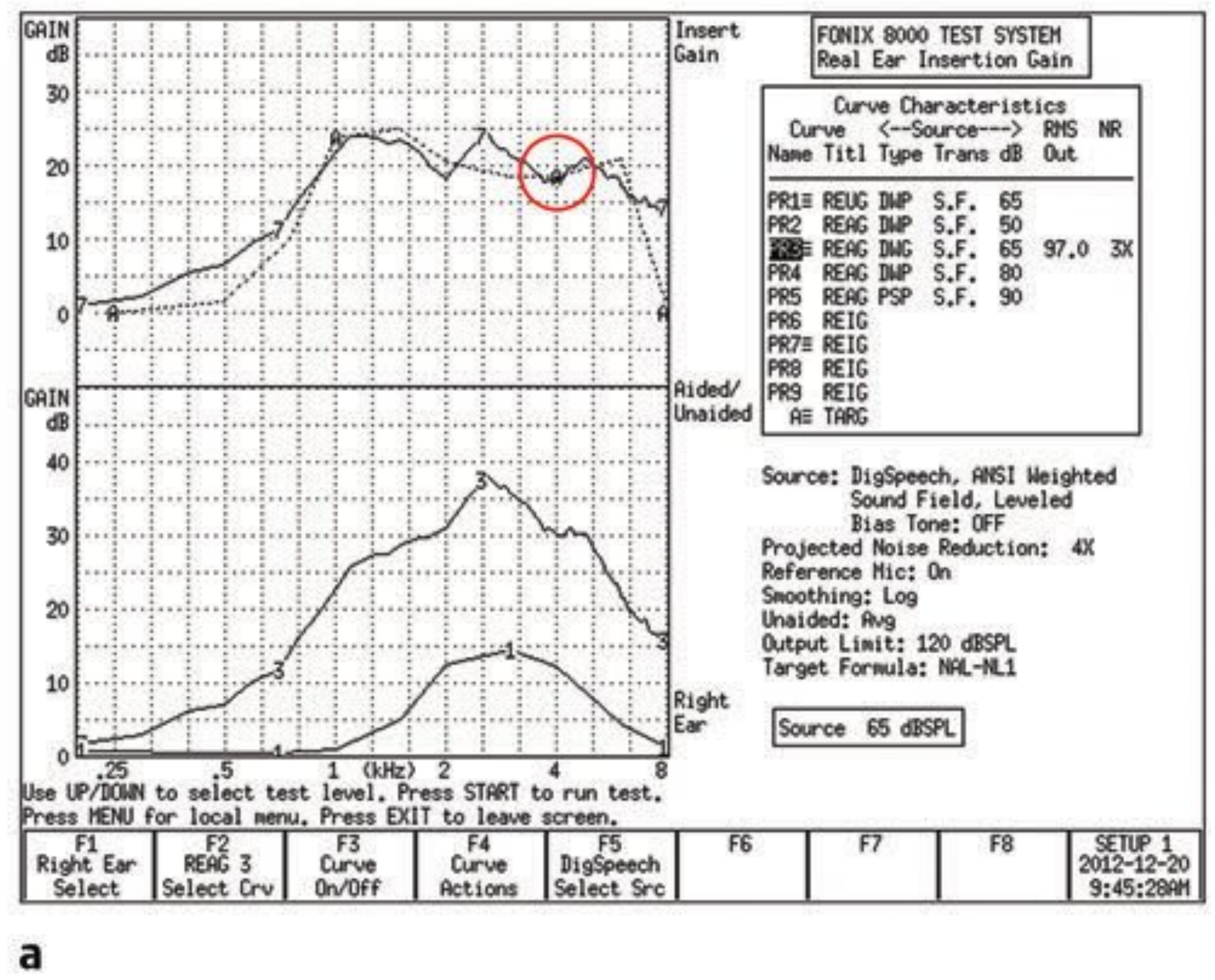
Status: Not OK (Measured) — In-situ vent effect: 4.0

b

Status: OK (Measured) — In-situ vent effect: 4.0

Status: OK (Measured) — In-situ vent effect: 4.0

Fig. 60.2 Results from the Widex feedback test for domes (a) and custom earmolds (b). In (a) and (b), the curve on the left is for the right ear and the curve to the right is for the left ear.



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FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve    <--Source---> RMS  NR
Name Titl Type Trans dB  Out

PR1≡ REUG DWP  S.F.  65
PR2  REAG DWP  S.F.  50
PR3≡ REAG DWG  S.F.  65  97.0  3X
PR4  REAG DWP  S.F.  80
PR5  REAG PSP  S.F.  90
PR6  REIG
PR7≡ REIG
PR8  REIG
PR9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Avg
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.
```

```
FONIX 8000 TEST SYSTEM
Real Ear Insertion Gain

      Curve Characteristics
  Curve    <--Source---> RMS  NR
Name Titl Type Trans dB  Out

PL1≡ REUG DWG  S.F.  65
PL2  REAG DWP  S.F.  50
PL3≡ REAG DWG  S.F.  65  93.9  4X
PL4  REAG DWP  S.F.  80
PL5  REAG PSP  S.F.  90
PL6  REIG
PL7≡ REIG
PL8  REIG
PL9  REIG
  A≡ TARG

Source: DigSpeech, ANSI Weighted
        Sound Field, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Reference Mic: On
Smoothing: Log
Unaided: Avg
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source  65 dBSPL

Use UP/DOWN to select test level. Press START to run test.
Press MENU for local menu. Press EXIT to leave screen.
```

Fig. 60.3 Measured real ear insertion gain (REIG) (curve 7) and prescribed REIG (curve A) for the right (a) and left (b) ears using a custom earmold.

reported that the fit was far more comfortable than what he had experienced over the previous 2 weeks using the domes.

60.3 Questions to the Reader

1. What is judged a significant decrease in hearing to warrant referral to an otologist?
2. Is it better to recommend referral to a general otolaryngologist or to an otologist in these cases?
3. The patient did not report tinnitus in either ear, and the results of reflex thresholds and reflex decay were consistent with cochlear involvement. Do you think a referral to an otologist was necessary?
4. What is the timeframe for a current audiogram to be completed before an audiologist would be comfortable going forward with fitting hearing aids.
5. When would you consider using powder and liquid material instead of silicone to make impressions of the ear canal?
6. What corrections, if any, should be applied to prescribe NAL-NL1?

60.4 Discussion of Questions to the Reader

1. What is judged a significant decrease in hearing to warrant referral to an otologist?
   At our clinic, a decrease in hearing level of 10 dB or greater at more than one frequency in either ear from the previous audiogram is considered significant. In addition, a decrease in a WRS is considered significant if the WRS at the follow-up visit exceeds the previous WRS using the binomial distribution. To determine this, the audiologist would look at Table 1 from Carney and Schlauch to compare the follow-up WRS to the previous WRS. Table 1 from Carney and Schlauch is placed at each audiometer in the authors' clinic for easy retrieval and interpretation. For example, in this case, the patient's previous WRS in the right ear was 92%, and the WRS for the follow-up visit was 76%. Using the 50-word list column in Table 1, the follow-up WRS must be poorer than 80% to be considered a significant decrease ($p < 0.05$). In this case, the follow-up WRS was 76%; thus the decrease in WRS was judged significant.
2. Is it better to recommend referral to a general otolaryngologist or to an otologist in these cases?
   The authors believe a referral to an otologist is more appropriate than a referral to a general otolaryngologist if there is concern about the integrity of any segment of the auditory system.
3. The patient did not report tinnitus in either ear, and the results of immittance, reflex thresholds, and reflex decay were consistent with cochlear involvement. Do you think a referral to an otologist was necessary?
   Yes, due to the significant decrease in hearing levels and WRS. In addition to making a recommendation to the patient's primary physician for a referral to an otologist, the audiologists also recommended that auditory brainstem response (ABR) testing be completed. The otologist instead ordered an MRI scan, the results of which were normal. In recent years, it has become increasingly common for some otologists to forgo the ABR and order an MRI scan.
4. What is the timeframe for a current audiogram to be completed before an audiologist would be comfortable going forward with fitting hearing aids?
   When fitting hearing aids, an accurate audiological examination must be completed no more than 6 months from the time the hearing aids are dispensed. In addition, the patient must obtain medical clearance for amplification or have signed a waiver (if an adult). At the authors' clinic, the audiologists strongly recommend a signed form for medical clearance and do not promote the signing of a waiver.
5. When would you consider using powder and liquid material instead of silicone to make impressions of the ear canal?
   This is a very important consideration if the patient is using any blood-thinning medication or has an autoimmune

disorder such as diabetes then powder and liquid, instead of silicone material should be used.

6. What corrections, if any, should be applied to prescribed NAL-NL1?

   It has been reported that 70 to 80% of hearing aids dispensed in the United States are completed without verification using REMs. For most of these, the audiologist or hearing aid dispenser simply downloads the manufacturer's First-Fit to the hearing aids. There is abundant research reporting that the manufacturer First-Fit will usually underamplify the high frequencies that are so important for speech recognition. For the remaining 20 to 30% of audiologists or hearing aid dispensers who do verify hearing aid fittings using REMs, most do not correct the REIG, REAR, or real ear saturation response (RESR) targets for the type of fitting (monaural or bilateral) or number of channels of signal processing. The first correction is for binaural summation. When applied, this correction reduces the REIG or REAR target by approximately 5 dB in each ear. The second correction is for channel summation, and this reduces the RESR target by 3 to 8 dB depending upon the number of channels of signal processing.

60.5 Outcome

The patient was very pleased with the performance of his hearing aids, remote control, and custom earmolds. He reported via an outcome measure that his hearing aids provided significant benefit, and he was very satisfied with the comfort and retention provided by the custom earmolds.

60.6 Key Points

1. Whenever possible, counsel patients on the advantages of custom earmolds and have the patient consider purchase of custom earmolds when fitting hearing aids coupled to the ear via thin-tubes or RICs.
2. It is good practice for audiologists to have a custom earmold (open and closed) made to fit their own ear to demonstrate to patients how a thin-tube or RIC hearing aid will look when coupled to the ear. Most hearing aid manufacturers will provide these at no charge if the audiologist sends an impression.
3. Develop a best practice guideline for your clinic to include the following:
   a) Use of coupler measures to verify the performance of all new and repaired hearing aids arriving at your clinic.
   b) Use of real ear measures to verify the performance to a validated prescriptive target of all hearing aids dispensed in your clinic. Do not use manufacturer First-Fit.
   c) Use of validation (outcome) measures to verify that the subjective preferences of all hearing aids dispensed in your clinic meet the expectations of your patients in satisfaction, benefit, and/or improved quality of life.
   d) Use full lists and recorded speech material when evaluating WRSs. Do not use half lists or monitored live voice.
   e) Establish protocols for when follow-up audiological examinations indicate significant changes in hearing and what recommendations should be made.

Suggested Reading

Carney E, Schlauch RS. Critical difference table for word recognition testing derived using computer simulation. J Speech Lang Hear Res 2007; 50: 1203–1209

Kuk F, Baekgaard L. Hearing aid selection and BTEs: choosing among various "open ear" and "receiver-in-the-canal options. Hear Rev 2008; 15: 22,24,26,31–32,36

Thornton AR, Raffin MJ. Speech-discrimination scores modeled as a binomial variable. J Speech Hear Res 1978; 21: 507–518

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Part 5

Cochlear Implants

61 Revisiting Cochlear Implant Candidacy 266

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64 Novel Use of Speech Recognition Testing for Counseling Cochlear Implant Recipients 278

61 Revisiting Cochlear Implant Candidacy

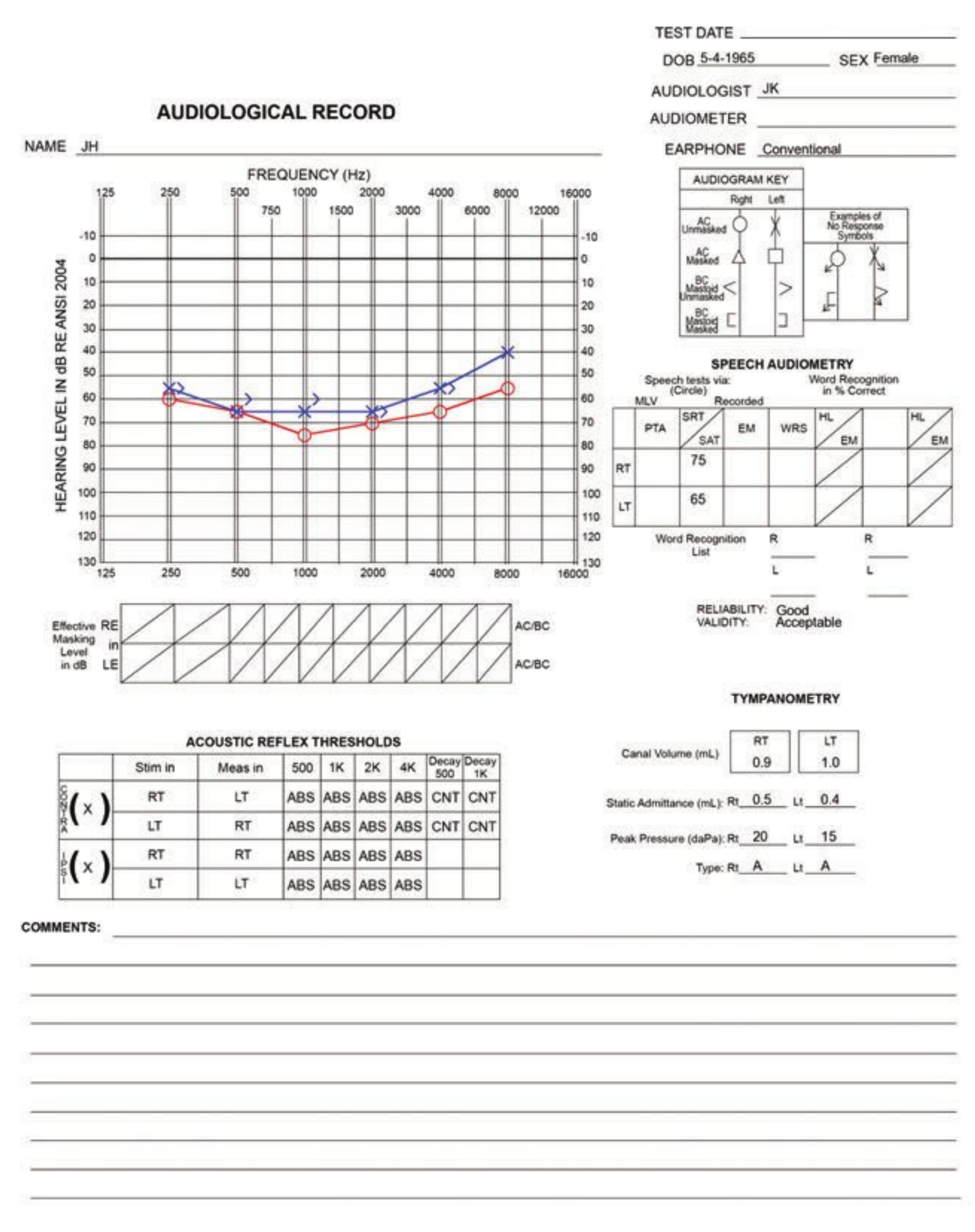
Jordan King and John L Dornhoffer

A 48-year-old female with a bilateral moderately severe sensorineural hearing loss undergoes cochlear implantation.

61.1 Clinical History and Description

JH is a 48-year-old female with a flat, moderately severe sensorineural hearing loss who was referred to evaluate her candidacy for a cochlear implant (CI) due to a recent decrease in hearing thresholds of 0 to 5 dB across all audiometric test frequencies in the right ear and 5 to 10 dB across all audiometric frequencies in the left ear. This threshold shift was accompanied by a significant decrease in word recognition scores (WRSs). The WRS for the right ear decreased from 84 to 60%, and the WRS for the left ear decreased from 78 to 36% Initial review of previous audiological examinations indicated more residual hearing than is typically allowable for candidates for a CI (Fig. 61.1) where JH's audiological evaluation revealed a moderate to moderately severe sensorineural hearing loss that is flat in configuration. Immittance audiometry revealed normal tympanograms and absent acoustic reflex thresholds (ARTs) to contralateral and ipsilateral stimulation at 500 to 1000 Hz. Typically, candidacy for a CI requires the patient to have a moderate to profound hearing loss. It was, however, decided to evaluate JH to determine potential benefit from amplification that could provide greater gain and output and performance than JH was receiving from her current hearing aids.

JH attended her initial appointment with her spouse and reported she was retired and spent a great deal of time at home with friends because her husband was often away on work related travel. JH reported increasing difficulty functioning without her spouse present because she often relied upon him to "translate" group conversations for her. JH was active in various social groups, including regular group camping outings, and



TEST DATE

DOB 5-4-1965 SEX Female

AUDIOLOGIST JK

AUDIOMETER

EARPHONE Conventional

AUDIOLOGICAL RECORD

NAME JH

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		75					
LT		65					

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.9	1.0

Static Admittance (mL): Rt 0.5 Lt 0.4

Peak Pressure (daPa): Rt 20 Lt 15

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	ABS	ABS	CNT	CNT
	LT	RT	ABS	ABS	ABS	ABS	CNT	CNT
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS:

Fig. 61.1 JH's preoperative audiogram.

she also served as a volunteer fire fighter in her community. JH reported that many of these activities were becoming difficult for her due to her hearing loss.

JH was initially diagnosed with hearing loss at age 27 years, and the hearing loss was reported as being progressive. JH reported wearing bilateral hearing aids since the time of her diagnosis. Her current hearing aids were bilateral ReSound in-the-ear (ITE) hearing aids (ReSound, Bloomington, MN) that were purchased 2 years ago. She and her spouse reported that she had “good hearing days and bad hearing days.” JH had graduated from college and has a very positive disposition.

61.2 Audiological Testing

A CI evaluation protocol at many clinics consists of pure-tone air and bone conduction thresholds, speech recognition thresholds (SRTs), word recognition scores (WRSs), immittance audiometry (i.e., tympanometry and acoustic reflex thresholds [ARTs]), and otoacoustic emissions (OAE) screening. Also included is a hearing aid evaluation (HAE) if a patient does not have amplification, hearing aid verification if a patient has amplification, followed by a loaner hearing aid fitting if indicated. The verification process consists of bilateral and monaural aided sound-field thresholds and bilateral and monaural aided speech recognition scores. These measures were completed with JH during four clinic visits over 4 months. JH consistently presented with normal tympanograms bilaterally, indicative of normal middle ear function. Ipsilateral and contralateral ARTs at 500, 1000, 2000, and 4000 Hz as well as distortion product OAEs were absent bilaterally, which is indicative of abnormal inner ear function and consistent with the magnitude of hearing loss indicated during behavioral testing.

After obtaining audiometric thresholds using insert earphones at 250, 500, 1000, 2000, 4000, and 8000 Hz, hearing aid verification of JH’s current hearing aids using real ear aided response (REAR) speech mapping was completed. Both hearing aids were verified to be underamplifying to the adult desired sensation level (DSL) prescriptive target for her hearing loss. Due to the patient’s desire to leave her hearing aid settings unchanged and be fit with new hearing aids, no attempt was made to adjust JH’s personal hearing aid settings. Earmold impressions were obtained bilaterally and new earmolds were ordered. JH was subsequently fit with bilateral loaner Phonak Extra 411 behind-the-ear (BTE) hearing aids (Phonak U.S., Warrenville, IL) using temporary Comply foam tips until her custom earmolds arrived. During her first and second clinic visits, JH’s loaner Phonak Extra 411 BTE hearing aids were adjusted until the two hearing aids appropriately met adult DSL targets for her hearing loss, and she reported the settings to be comfortable. JH was pleased with the loaner hearing aid fitting, but upon JH’s return for her third visit, JH and her spouse reported no noticeable functional improvement. Despite the hearing aids being appropriately programmed to the DSL target, the desired aided sound-field threshold of 25 dB HL or better at all test frequencies to frequency-modulated (FM) pure-tones presented at 0° azimuth could not be achieved for bilateral (0–0) and best monaural (X-X) aided sound-field testing (Fig. 61.2).

JH was provided a 1-month period of adjustment in order to acclimatize to the new settings before aided WRSs were measured at her next visit. This testing was completed in order to evaluate JH’s functional benefit provided by the loaner Phonak Extra 411 BTE hearing aids. The current speech recognition test battery in many CI centers includes AzBio sentences and consonant-nucleus-consonant (CNC) words at a minimum presentation level of 50 dB sound pressure level (SPL). JH’s evaluation, however, occurred before the clinic revised the test battery protocol and JH was administered the hearing in noise test (HINT) and CNC word test. For the purposes of qualifying for a CI the HINT was administered in quiet and scored in percent correct. Using speech recognition tests, current criteria for successful candidates for CI are 60% or poorer in the best aided condition, which is typically defined as using bilateral amplification with a score of 50%or poorer in the ear to be implanted. JH’s scores were within the CI candidacy range when tested in the bilateral aided condition at 50 dB HL (59%for HINT at 50 dB HL and 32%for CNC at 50 dB HL) and in the monaural conditions at 60 dB HL (51% for the right aided and 30% for the left aided) (Table 61.1)

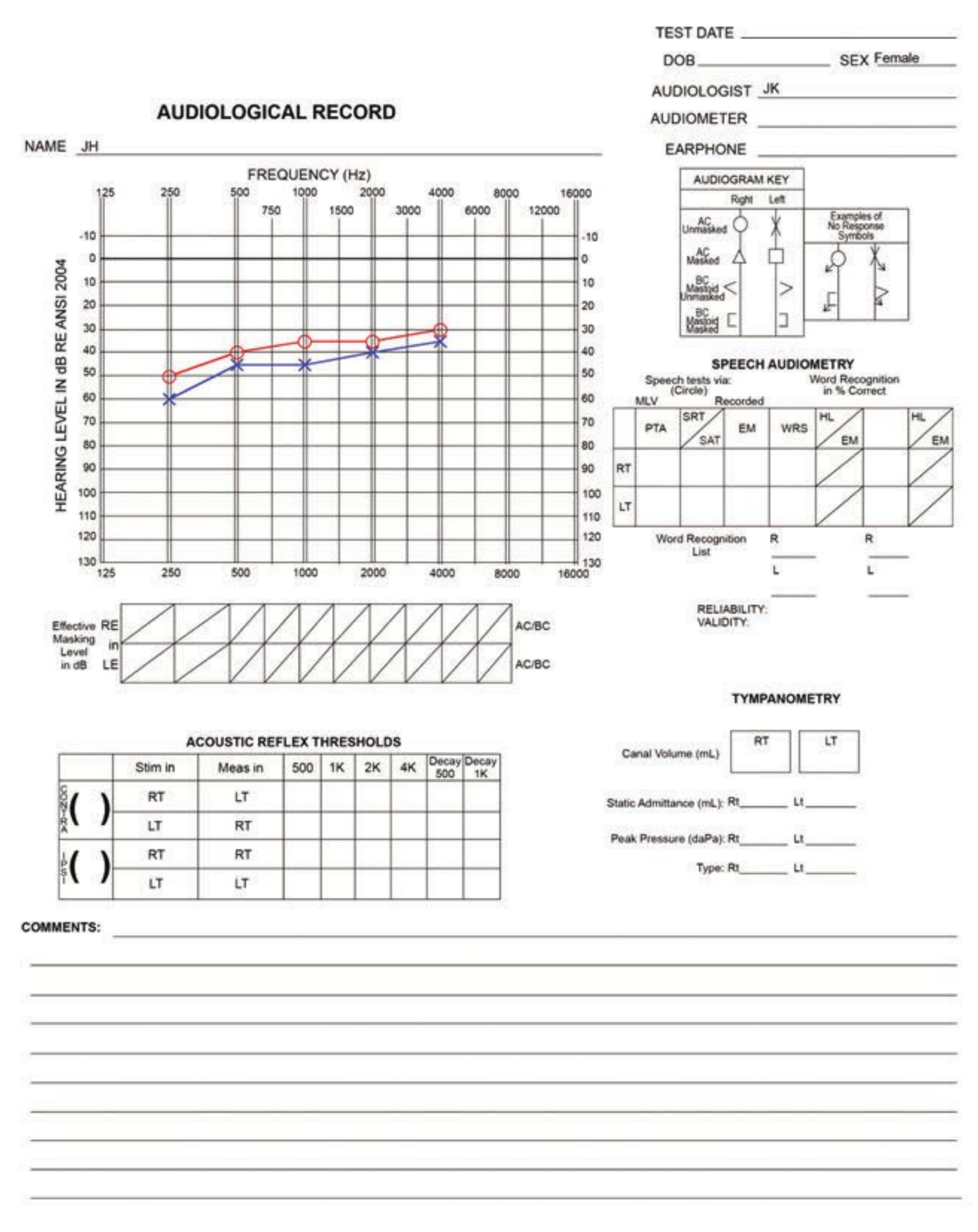
61.3 Questions to the Reader

1. What additional testing, if any, might be recommended based on JH’s reported case history and the measured aided sound-field thresholds?
2. What are some considerations when implanting a patient “outside” the recommended device criteria (i.e., “off-label”)?
3. Does a patient’s funding source determine his or her CI candidacy, particularly when the patient does not meet all the candidacy criteria?

61.4 Discussion of Questions to the Reader

1. What additional testing, if any, might be recommended based on JH’s reported case history and the measured aided sound-field thresholds?

   In this case report, JH’s audiologist lacked a clear explanation for JH’s inability to achieve the desired prescribed aided sound-field thresholds (i.e., 25 dB HL or better) or the desired aided speech recognition improvement given JH’s magnitude of hearing loss, which was felt to be aidable. Given JH and her spouse’s report of “good and bad hearing days,” the audiologist elected to recommend auditory brainstem response (ABR) testing. ABR testing was used to rule out auditory neuropathy spectrum disorder (ANSD). JH’s WRSs were within the criteria for CI candidacy; however, JH’s audiometric thresholds were not. A moderate sloping to profound hearing loss is the most widely accepted minimum magnitude and configuration of hearing loss for candidacy for a CI, and JH presented with significantly better residual hearing. The audiologist needed to obtain a greater understanding of the etiology of JH’s hearing loss in order to counsel JH on her possible outcome related to CI use. ABR results indicated a waveform I, III, V complex for JH’s right and left ears at threshold and suprathreshold levels for condensation and rarefaction stimulation. This finding ruled out ANSD as a possible etiology of JH’s hearing loss.



AUDIOLOGICAL RECORD

NAME JH

SEX Female

AUDIOLOGIST JK

Fig. 61.2 JH's preoperative aided sound-field thresholds with hearing aids for bilateral (0–0) and best monaural (X-X) fitting.

2. What are some considerations when implanting a patient "outside" the recommended device criteria (i.e., "off-label")?

   The primary concern was the possibility of permanent loss of JH's residual hearing. Currently, CI manufacturers and surgeons are united in efforts to develop methods to surgically implant CIs while preserving cochlear structures and maintaining residual hearing. If the recommendation for a CI was agreed to by JH, she would undergo CI surgery at a time when the possibility of residual hearing preservation was not favorable. Nontraumatic surgical techniques and nontraumatic electrode arrays continue to be developed and perfected. At the time JH would receive her CI, however, little success had been reported regarding hearing preservation. In addition to permanent loss of acoustic hearing, JH's expectations and her readiness for the CI were very important. A significant amount of time was spent counseling JH to the high variability in performance among CI recipients. Although a clinician was able to counsel JH that "typical" CI recipients perform better than she did with her amplification on aided speech recognition, JH's performance with her hearing aids was poorer than expected given the magnitude of her hearing loss. This was concerning and was brought to JH's attention as a possible contraindication that, even with improved audibility, the origin of her hearing loss might prevent a significant improvement in speech clarity with her CI. JH stated she felt she had "nothing to lose" and that losing her residual hearing in one ear was a risk she was "willing to take" if there was any chance of improving her hearing.

Table 61.1 JH's preoperative speech perception scores

	HINT at 60 dB HL	HINT at 50 dB HL	CNC at 50 dB HL
Binaural aided	85%	59%	32%
Right aided	51%	DNT	DNT
Left aided	30%	DNT	DNT

Abbreviations: CNC, consonant-nucleus-consonant; DNT, did not test; HINT, hearing in noise test.

3. Does a patient's funding source determine his/her CI candidacy, particularly when the patient does not meet all the candidacy criteria?

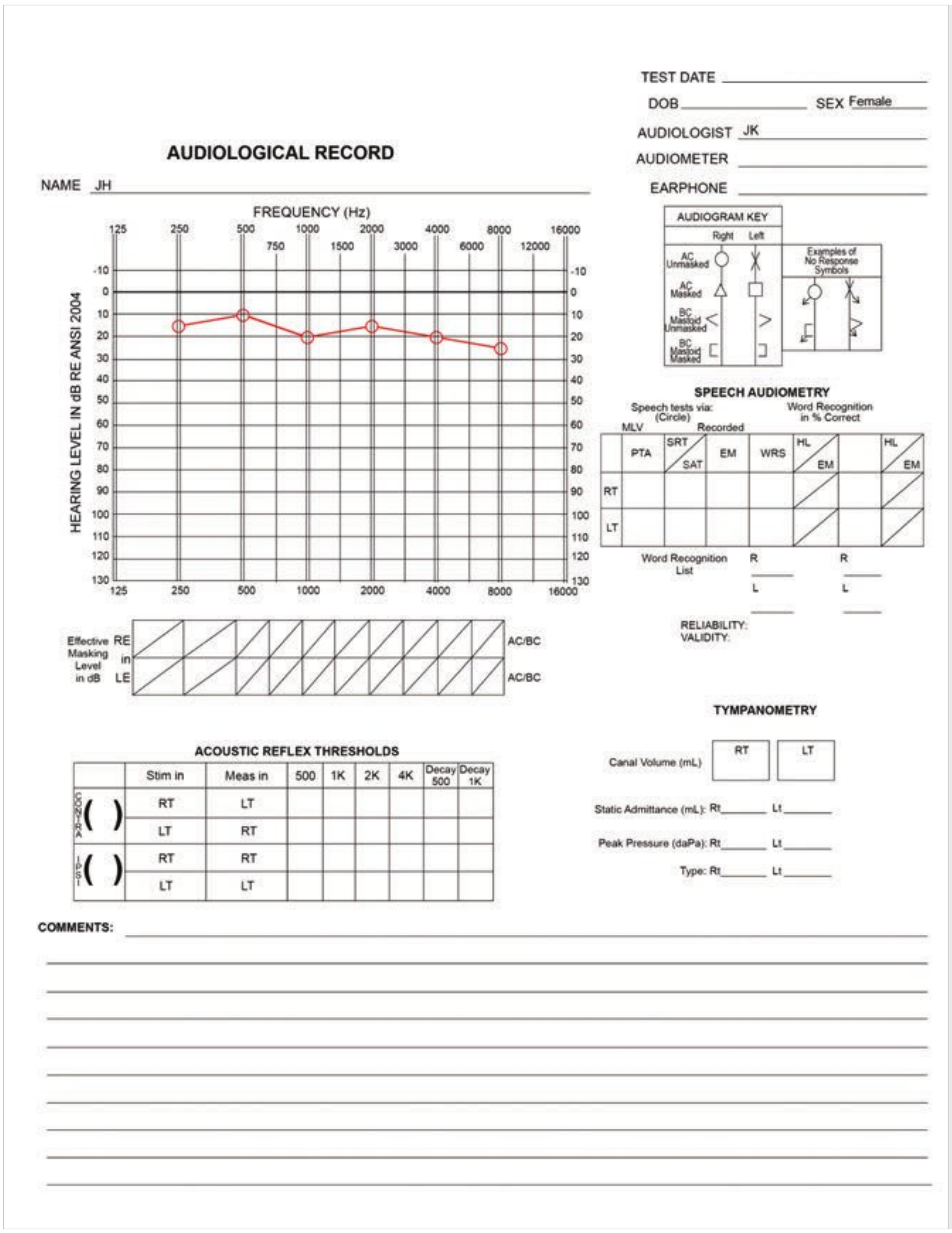
   Yes, different funding sources (i.e., private insurance, Medicare, Medicaid) have different candidacy guidelines that must be observed in order to receive the best possible reimbursement. It is the practitioner's advantage to strictly follow candidacy guidelines per the patient's funding source in order to obtain reimbursement and to avoid any unexpected "out of pocket" expenses on the part of the patient. JH had private insurance and was able to obtain approval from her insurance to receive the CI even though her case was considered off-label. It should be noted that not all private insurance policies have the same benefits, and that similar cases may have a different determination from the insurance company per the patient's policy.

61.5 Diagnosis and Recommended Treatment

JH's performance had not improved with an optimized hearing aid fitting. JH appeared to clearly understand the potential benefits and risks involved with proceeding with the CI, and she had a very strong desire to do so. JH was scheduled for CI surgery to her left ear. Although JH's left ear had slightly better hearing thresholds, JH had a strong preference to be implanted on her left ear based on hearing history and the dominance of her right ear for use of the telephone. JH elected to proceed with implanting the left ear, and she was implanted with the Advanced Bionics Hi Res 90K (Advanced Bionics LLC, Valencia, CA).

61.6 Outcome

Following activation of JH's left CI, aided sound-field thresholds with the CI only and using her hearing aid turned off to be used as an ear plug indicated an improvement when compared with preoperative aided sound-field thresholds with her hearing aids (Fig. 61.3). At the time of JH's most recent examination, JH was 3 years and 7 months removed from activation of her CI and her aided sound-field thresholds remained stable. JH continues to exhibit significant benefit from her CI with regard to speech recognition abilities (HINT-Quiet; HINT+10 signal to noise; CNC words) compared to preoperative measurements with her hearing aids (Fig. 61.4). A unique component of a good adult CI program is a required aural rehabilitation (AR) program in the form of auditory-based therapy administered by a speech language pathologist (SLP) for a minimum of 2 months postactivation. Longer enrollment times for AR services are



AUDIOLOGICAL RECORD

NAME JH

TEST DATE

DOB

SEX Female

AUDIOLOGIST JK

AUDIOMETER

EARPHONE

COMMENTS:

Fig. 61.3 JH's postoperative aided sound-field thresholds with cochlear implant.

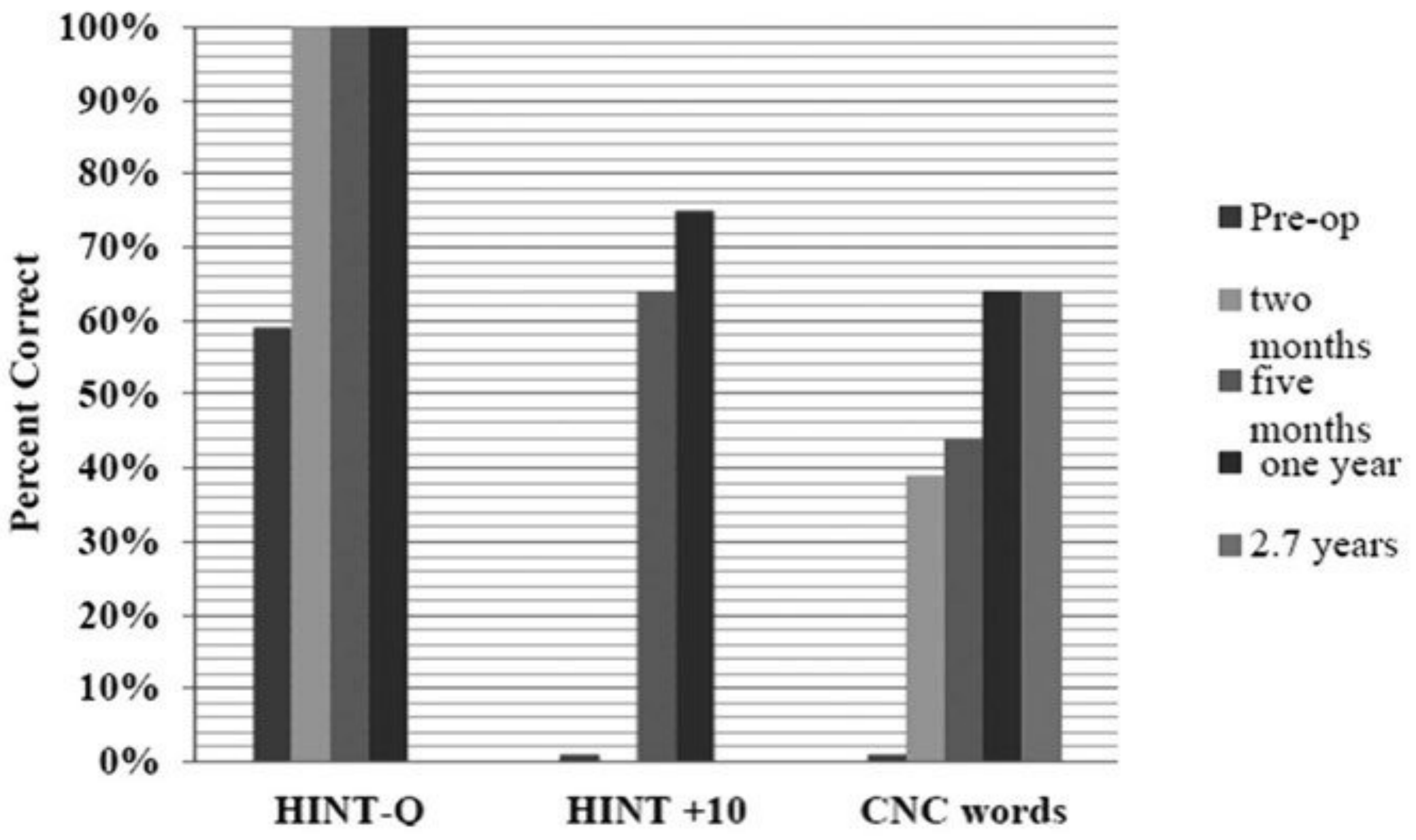


Fig. 61.4 JH's speech recognition scores preoperatively (preop) and at 2 months, 5 months, 1 year, and 2.7 years.

optional, and at times favorable, given a patient's progress and personal goals. The feedback provided from the SLP through the required therapy assisted the audiologist who was programming JH's CI to more easily achieve mapping optimization and helped JH adjust more quickly to her CI. In an e-mail from JH received approximately 6 weeks postactivation she wrote, "Out to lunch, my friend from New Jersey spoke very fast and she forgot to turn toward me. She sat across from me instead of beside me, but I decided to see how it went. Well . . . not only could I keep up with her conversation, but I could eat and hit my mouth because I could watch my fork rather than read her lips!!! I am continually blown away with what I can hear with my CI."

61.7 Key Points

1. Criteria have broadened since CIs were approved in 1985. A wider range of recipients are now able to enjoy higher levels of success with CIs than is often possible with hearing aids.
2. Functional ability cannot always be easily or accurately estimated based only on audiometric thresholds. In many cases, where significant residual hearing is present, it may be beneficial to enroll patients in preoperative auditory-based therapy. This therapy helps to ensure that patients are able to use their residual hearing to the best of their ability prior to electing to undergo an invasive procedure such as CI.
3. When evaluating a patient for CI, a comprehensive patient profile such as functional listening abilities in formal soundfield testing and real world listening should be taken into account.

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62 Clinical Management of a Patient with a Folded Electrode Array

Sarah O. Holbert, Michael J. Cevette, Linsey Scheibler, David Barrs, and Sarah Shepherd

A 78-year-old man undergoing reimplantation of his hybrid electrode was clinically evaluated following loss of residual hearing in his right ear. At the time of the assessment, the left ear standard array cochlear implant (CI) revealed an electrode “fold-over” (i.e., electrode had folded back on itself).

62.1 Clinical History and Description

DR was a 78-year-old man who presented to a clinic in October 2011 for reprogramming his right Cochlear Corporation Hybrid electrode and left Freedom Contour electrode (Cochlear, Centennial, CO). He reported receiving the Cochlear Corporation S8 Hybrid array (10 mm with six electrodes) in 2007 as part of a Food and Drug Administration (FDA) trial. For the duration of the trial he used a combination of hearing aid and CI (bimodal fitting) in his right ear. He later discontinued use of the hearing aid in his right ear after completion of the trial because of a nearly complete loss of his residual low-frequency hearing. DR then received a standard electrode array in 2008 in his left ear at a different institution. At that time, he was also offered the option of reimplantation with a standard electrode array in his right ear. He declined, however, because of a difficult post-surgical recovery due to dizziness and disequilibrium following the initial implantation of his left ear.

At the time of his evaluation at a clinic in 2011 for reprogramming of his CIs, he reported limited speech recognition from his right CI. His left CI was providing satisfactory benefit, aside from a persistent “hollow” sound quality. DR reported a history of gradual hearing loss that progressed over the past 25 years with consistent use of bilateral hearing aids for 23 years. In addition, DR reported that, although not bothersome, he heard tinnitus in the form of various noises including music, voices, and an engine noise bilaterally. He reported no other otologic symptoms.

62.2 Audiological Testing

Results of DR’s comprehensive audiological examination revealed a profound sensorineural hearing loss (SNHL) bilaterally with measurable hearing at 500 to 750 Hz (105–110 dB HL) in the right ear only. A CI-assisted aided sound-field threshold evaluation using frequency-modulated (FM) warble tones revealed thresholds in the 40 to 95 dB HL range for his right hybrid CI and thresholds at 15 to 30 dB HL for his left CI. Speech recognition testing using AzBio sentences presented at 60 dB SPL (A-weighting) in quiet revealed speech recognition of 10% in the right CI only, 76% in the left CI only, and 76% with bilateral CIs.

62.3 Questions to the Reader

1. Describe the candidacy criteria used for the FDA hybrid trials.
2. What is the incidence of dizziness and disequilibrium following implantation, and what tests might be completed to evaluate this condition? Why would such testing be important in considering which ear, or both, to reimplant?

62.4 Discussion of Questions to the Reader

1. Describe the candidacy criteria used for the FDA hybrid trials.
   FDA inclusionary criteria for implantation of hybrid devices from Cochlear Corporation (including the S8 device, S12, and L24) are as follows:
   a) ≥18 years at the time of implantation
   b) Severe to profound hearing loss above 1500 Hz with a threshold of 75 dB HL or greater from 2000 to 4000 Hz
   c) Low-frequency thresholds no poorer than 60 dB HL
   d) Consonant-nucleus-consonant (CNC) word recognition scores between 10 and 60% in the ear to be implanted
   e) CNC word recognition scores in the contralateral ear ≥ those in the ear to be implanted but not to exceed 80% (Gantz el al, 2009)
2. What is the incidence of dizziness and disequilibrium following implantation, and what tests might be done to evaluate this condition? Why would such testing be important in considering which ear, or both, to reimplant?
   Research suggests that approximately 40% of patients experience new-onset dizziness following cochlear implantation. The patient might be evaluated using computerized dynamic posturography (CDP), videonystagmography (VNG) including caloric stimulation, vestibular-evoked myogenic potentials (VEMPs), and rotary chair (RC). The vestibular battery would be used to evaluate vestibular function in each ear. If the patient only had vestibular function in one ear (as shown by a present VEMP and caloric response), a reimplantation in that ear could cause the patient to lose all remaining vestibular function and therefore be functionally impaired with his balance.

62.5 Additional Testing and Treatment

Due to DR’s apprehension to undergo a revision surgery for his right CI, programming was attempted to improve his aided sound-field thresholds and speech recognition. In addition to the typical programming measurements including programming threshold (T) and comfortable (C) levels, DR’s frequency allocation table (FAT), which defines the frequency range or bandwidth assigned to each active channel, was adjusted. DR preferred the sound quality with MAP settings using an upper cutoff frequency of 6000 Hz and a lower cutoff frequency of 70 Hz. Following these adjustments, his aided sound-field

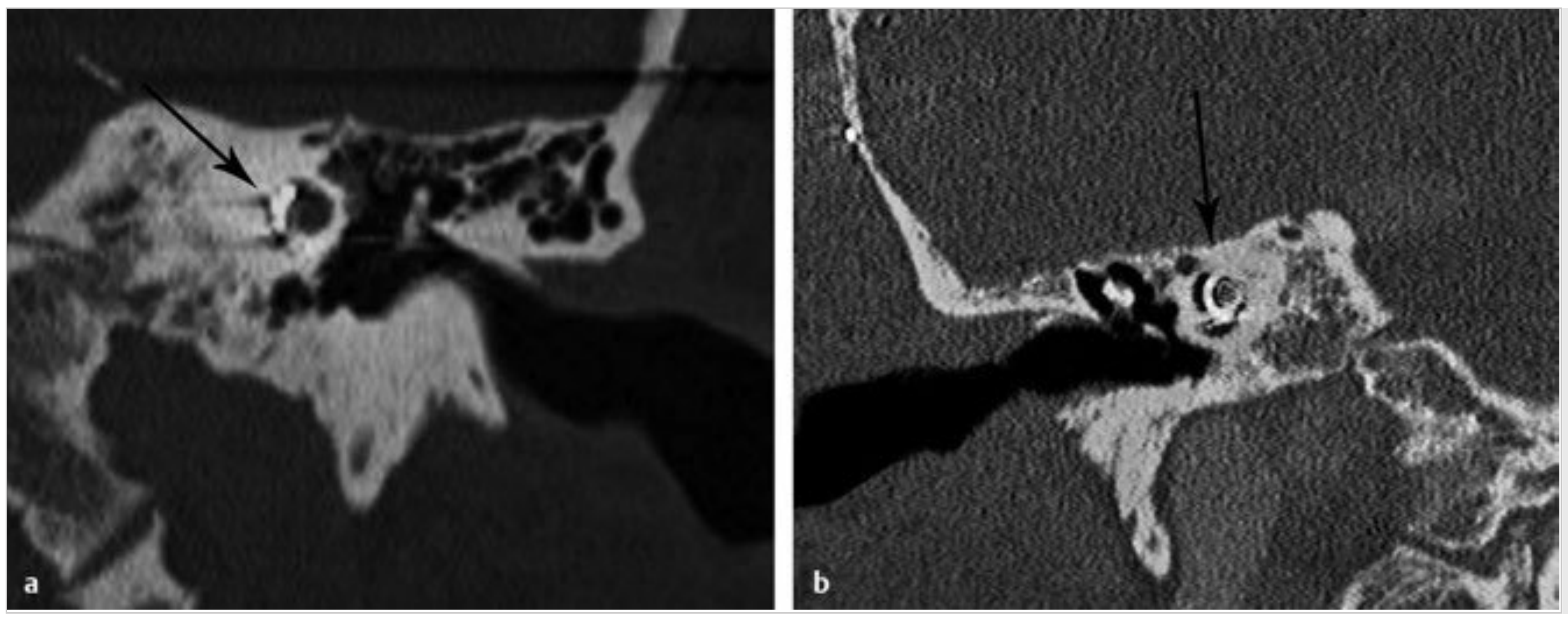

Fig. 62.1 (a) Computed tomographic (CT) scan showing fold-over electrode array. The arrow points to the ascending limb of the basal turn where the electrode had folded back on itself, instead of completing a full insertion of the basal turn. (b) CT scan showing full insertion of an electrode array. The arrow points to the CI leads extending through the basal turn of the cochlea.

thresholds improved to the expected levels at 250 to 3000 Hz. His thresholds at 4000 to 6000 Hz remained at 45 dB HL, and "no response," respectively. Speech recognition testing 1 month later revealed no significant change.

DR met with the otologist to discuss reimplantation. As part of his workup, a computed tomographic (CT) scan was ordered and reported good positioning of the short hybrid CI leads in the basal turn of the right cochlea. The CT, however, also showed a malpositioned CI electrode in the basal turn of the left cochlea. In the ascending limb of the basal turn, the electrode had folded back on itself, instead of completing a full insertion of the basal turn (Fig. 62.1). His vestibular assessment was normal bilaterally and included VNG and CDP.

62.6 Diagnosis and Recommended Treatment

Based on the results of his evaluation, reimplantation of the right ear using a standard electrode array was recommended. Dr was counseled that patients who are reimplanted with a standard array often experience improved speech recognition. A full electrode insertion of the right ear was achieved using a Cochlear Corporation Freedom Contour electrode. Placement was confirmed using a plain film X-ray. Neural response telemetry (NRT), an objective measure of each electrode's response to electrical stimulation, revealed responses across several electrodes with good morphology.

DR's new, right CI was activated 1 month postsurgery, and he noticed an immediate improvement in speech recognition and "fuller" sound quality than with his left CI. Aided sound-field thresholds using frequency-modulated pure-tones were 20 to 25 dB HL. Speech recognition testing revealed the right ear CI surpassed the left ear speech recognition at 6 months post reactivation. AzBio sentences in quiet revealed an improvement of 10 to 92% for the right CI, a score of 68% for his left CI, and a score of 96% with bilateral CIs (Fig. 62.2 and Fig. 62.3).

62.7 Additional Questions to the Reader

1. What type of testing can show electrode fold-over and what are the clinical signs that strongly correlate with fold-over?

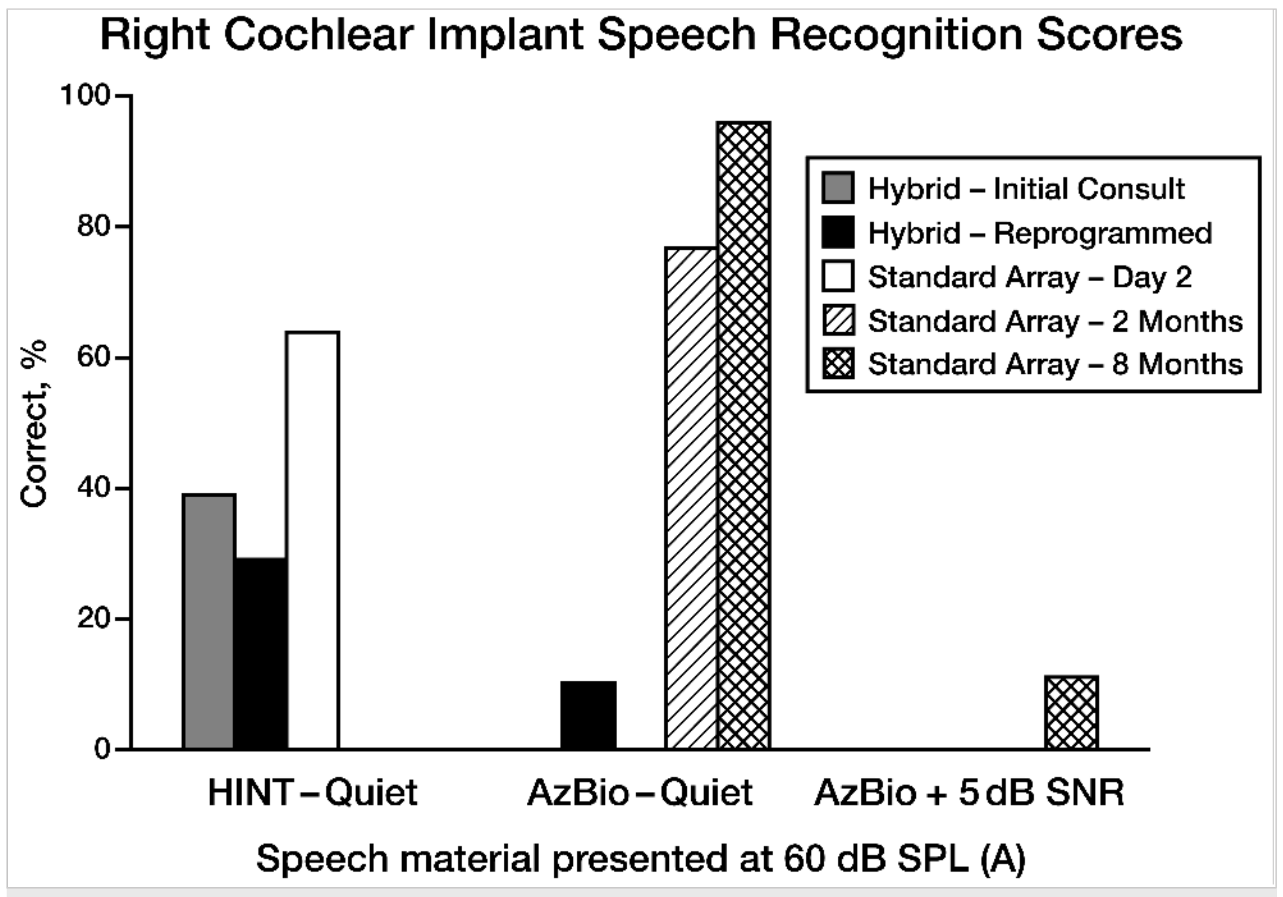

Fig. 62.2 Speech recognition testing for patient 6 months post reimplantation of a Cochlear Corporation right hybrid with standard electrode array and 5 years postimplantation of a left Cochlear Corporation standard electrode.

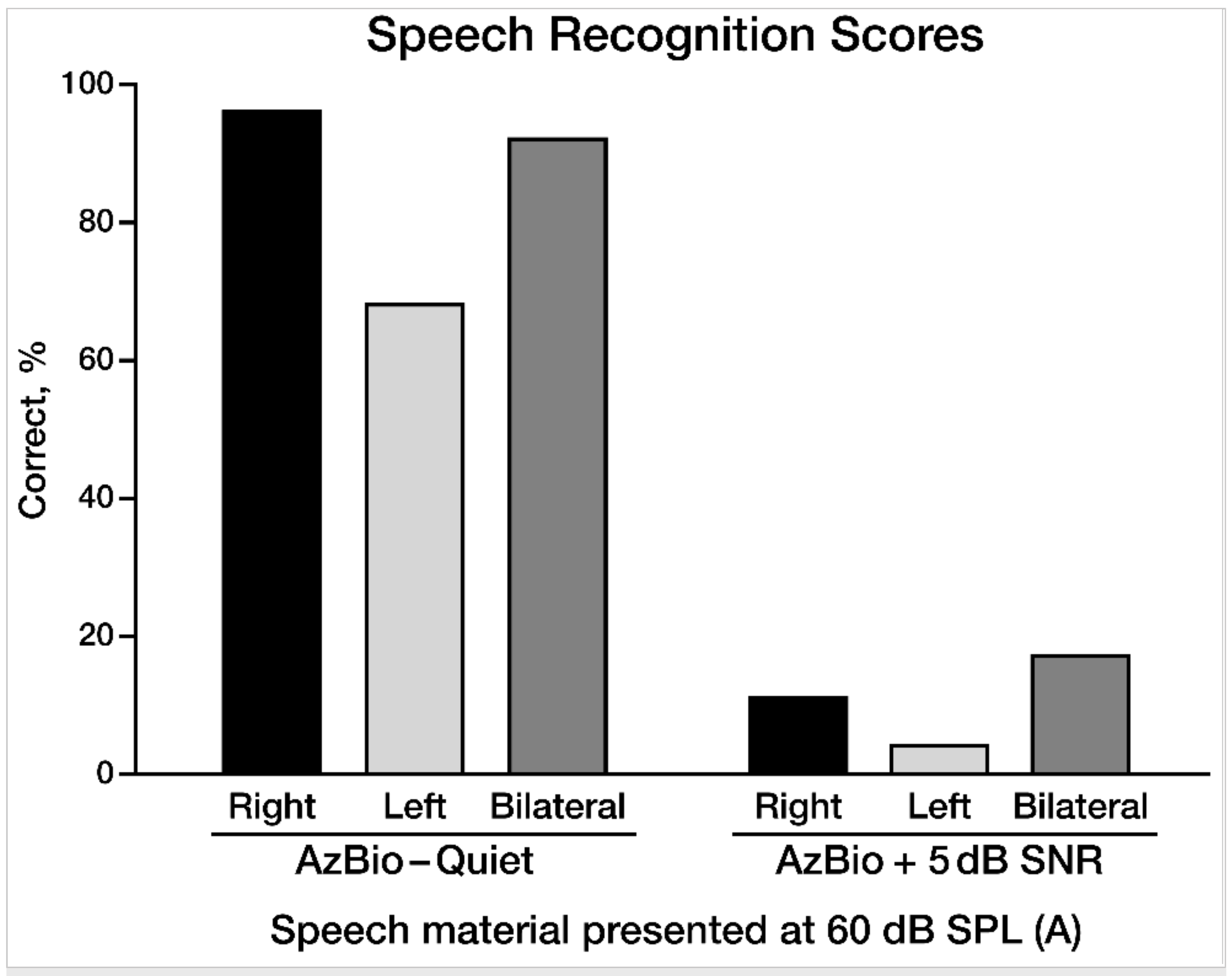

Fig. 62.3 Speech recognition testing of patient with Cochlear Corporation hybrid and reimplanted standard electrode array in right ear.

2. Despite the presence of an electrode fold-over, DR performs well with his left CI. What is considered average performance of a typical late deafened adult CI user? Should reimplantation of DR's left ear be considered?

62.8 Discussion of Additional Questions to the Reader

1. What type of testing can show electrode fold-over and are there clinical signs that strongly correlate with fold-over?
   A plain film X-ray and/or spread of excitation (SOE) testing are often the simplest methods to help identify electrode fold-over at surgery. A CT scan may also be used and is considered by many as the gold standard. In most facilities, however, it is not easily accessible in the operating room.
   DR did report a “hollow” sound quality; however, this can often be corrected with programming adjustments and does not necessarily correlate with an electrode placement issue. In this particular case, especially since DR reported a “fuller” and improved sound quality from the reimplanted right ear, the sound quality issue may have been an early indicator of the fold over. The most significant clinical sign, however, warranting investigation is poor speech recognitions scores.
2. Despite the presence of an electrode fold-over, DR performs well with his left CI. What is considered average performance of a typical late-deafened adult CI user? Should reimplantation 006Ff DR’s left ear be considered?
   Average performance for AzBio sentences is 70%correct in quiet. Despite the fold-over in DR’s left CI array, DR performed at an average level. Generally, speech recognition performance is a significant factor when considering reimplantation. Because DR is considered an average performer, it is questionable whether his left ear should be reimplanted. Patient preference, age, and risks of an additional surgery are all factors for consideration.

62.9 Key Points

1. Reimplantation is an option for patients with hybrid electrodes if they lose their residual low-frequency hearing.
2. Although DR performs well with both implants, electrode fold-over often impacts performance and perception of sound quality. Plain film X-rays, CT, and SOE testing are methods available for identifying electrode fold-over.

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63 Test Measures Matter

Sarah A. Sydlowski

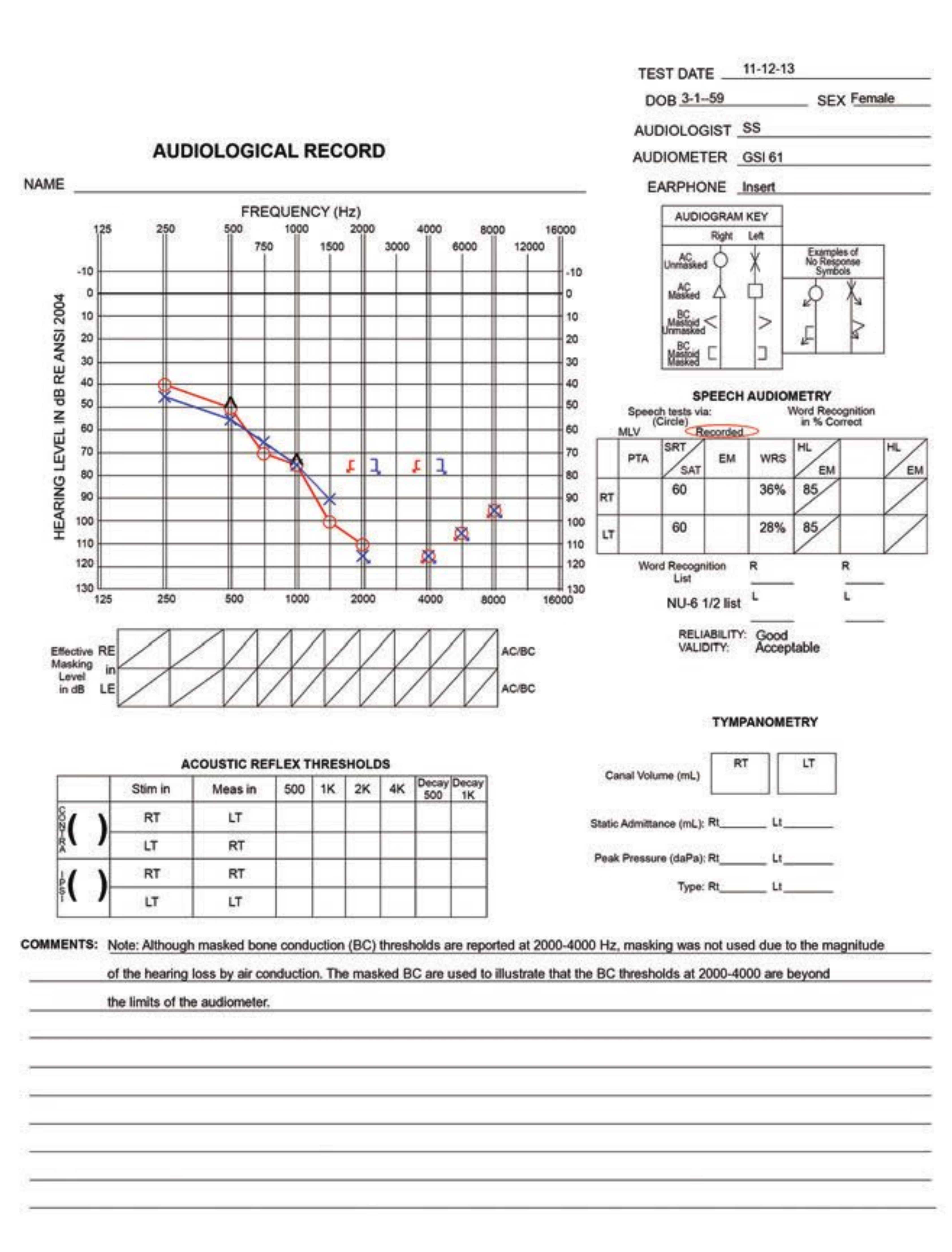
A patient's candidacy for a cochlear implant (CI) depends on the use of appropriate assessment measures.

63.1 Clinical History and Description

A 54-year-old female was referred for a cochlear implant (CI) evaluation. Her hearing loss reportedly began approximately 15 years ago. The etiology of her hearing loss is unknown but likely has a genetic component because her mother also had a similar pattern of hearing loss. She first began using bilateral hearing aids 12 years ago and has been consistently aided appropriately during that time. When she presented for her CI evaluation she was wearing bilateral Phonak Naida V UP hearing aids (Phonak U.S., Warrenville, IL). The performance of her hearing aids was verified to meet National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1) prescriptive targets for soft (55 dB SPL), average (65 dB SPL), and loud (75 dB SPL) input levels. She also used a Phonak ZoomLink transmitter and Phonak MyLink FM receiver. She reported significant difficulty, particularly in challenging listening environments such as background noise. The patient reported that her place of employment is very noisy, and her hearing loss is impacting her ability to successfully complete her job. She also reported use of closed captioning and wireless TVEars (TV Ears, Inc., Spring Valley, CA) for listening to the television. She can occasionally use the telephone but has started to rely more heavily on e-mail if the caller is not a close friend or family member.

63.2 Audiological Testing

At the time of her audiological examination (Fig. 63.1) her hearing loss was found to be a bilateral symmetrical sensorineural hearing loss (SNHL). The right ear revealed a mild SNHL



AUDIOLOGICAL RECORD

TEST DATE 11-12-13
DOB 3-1--59 SEX Female
AUDIOLOGIST SS
AUDIOMETER GSI 61
EARPHONE Insert
NAME

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		60		36%	85		
LT		60		28%	85		

Word Recognition List R ______ R ______

NU-6 1/2 list L ______ L ______

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)		

Static Admittance (mL): Rt_______ Lt_______
Peak Pressure (daPa): Rt_______ Lt_______
Type: Rt_______ Lt_______

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS: Note: Although masked bone conduction (BC) thresholds are reported at 2000-4000 Hz, masking was not used due to the magnitude of the hearing loss by air conduction. The masked BC are used to illustrate that the BC thresholds at 2000-4000 are beyond the limits of the audiometer.

Fig. 63.1 Results from the audiological examination. Unaided word recognition at 85 dB HL (most comfortable level): 36% right, 28% left

Table 63.1 Aided testing using HINT sentences

Test condition	In quiet
Right	69%
Left	64%
Bilateral	88%

at 250 Hz sharply sloping to a moderate to profound SNHL at 500 to 8000 Hz with no measurable hearing at 3000 to 8000 Hz. The left ear revealed a moderate SNHL at 250 Hz sharply sloping to a profound SNHL at 500 to 8000 Hz with no measurable hearing at 2000 to 8000 Hz. Her word recognition score (WRS), using the Northwestern University Auditory Test No. 6 (NU-6) word lists revealed very poor ability to recognize speech bilaterally. The patient was initially evaluated for CI candidacy using the Hearing in Noise Test (HINT) sentences presented in quiet from 0° azimuth per the Minimum Speech Test Battery (MSTB, 2003) protocol (Table 63.1). Her HINT scores were 69%, 64%, and 88% for the right, left, and bilateral aided conditions, respectively, indicating good aided performance provided by her hearing aids in a quiet listening condition.

The U.S. Food and Drug Administration (FDA) labels each CI manufacturer's device with specific candidacy criteria. Clinicians often use the more generalized criteria of 50% recognition of words in sentences in the aided condition for the ear to be implanted, and 60% for the contralateral ear and the bilaterally aided conditions. As already mentioned, this patient recognized 69% of words with her right hearing aid only, 64% with her left hearing aid only, and 88% with bilateral amplification. Because her speech recognition scores using HINT sentences exceeded FDA speech recognition criteria for a CI, the patient was informed that she was not a candidate for a CI.

63.3 Questions to the Reader

1. What are the current CI candidacy criteria for adults?
2. Why was this patient able to achieve such relatively high scores on HINT sentences despite her poor unaided WRSs?
3. Which additional tests, if any, should be administered?

63.4 Discussion of Questions to the Reader

1. What are the current CI candidacy criteria for adults?
   A CI is regulated by the FDA and insurance carriers. Each CI manufacturer's device candidacy guideline is labeled for its indicated use. Typically, the guidelines specify candidacy criteria as bilateral moderate to profound SNHL and reporting "limited benefit" from amplification. "Limited benefit" from amplification, historically, has been defined in the United States by using speech recognition scores when listening to open-set sentences in quiet under the best aided condition. The labeling for Cochlear™ Americas' CI is most commonly referenced in the literature and specifies ≤ 50% WRSs in the ear to be implanted and ≤ 60% using the best aided condition (Cochlear™ Americas package insert). In looking at the results in Table 63.1 it is clear that the patient in this case report does not meet those criteria.
2. Why was this patient able to achieve such relatively high scores on HINT sentences despite her poor unaided WRSs?
   HINT sentences consist of a single male talker speaking highly contextual sentences in a relatively slow, well-enunciated voice. Because the sentence presentations are slow, well-enunciated, and presented in quiet, this patient is able to use the context of the sentences to understand and repeat most of the words. Conversely, the NU-6 word lists to assess the patient's unaided WRS lack the context to help the patient ascertain the speaker's meaning even without complete audibility. HINT sentences were originally developed to be used in an adaptive presentation in a background of noise; however, most clinicians using HINT sentences for assessment of CI candidacy present the sentences in quiet. Gifford et al indicated that HINT sentences suffer from ceiling effects and may not be sufficiently challenging to capture CI candidates' and recipients' speech recognition capabilities.
3. Which additional tests, if any, should be administered?
   In order to better approximate the typical patient's real-world listening environment, it is important to incorporate test measures that use male and female talkers presented in quiet and noise using sentence and single-word materials. In 2011, the revised MSTB was developed and released. The revised MSTB includes (1) consonant-nucleus-consonant (CNC) monosyllables, (2) Bamford-Knowles-Bench Speech-in-Noise (BKB-SIN) test, and (3) AzBio sentences presented in quiet and in a fixed-level multitalker background noise. Using the new MSTB should produce patient performance that does not approach ceiling limits and offers more ecologically valid stimuli.

63.5 Additional Testing

A CI evaluation was conducted using the revised MSTB presented in sound field under the right-aided, left-aided, and bilaterally aided conditions. The results of the MSTB for this patient are summarized in Table 63.2 for the AzBio sentences, CNC words, and BKB-SIN words.

Table 63.2 Aided testing using the revised Minimum Speech Test Battery

Aided AzBio sentences		
Test condition	In quiet	
Right	45%	
Left	36%	
Bilateral	54%	
Aided CNC words		
Test condition	Words	Phonemes
Right	20%	47%
Left	0%	21%
Bilateral	4%	33%
Aided BKB-SIN		
Test condition	SNR loss	Degree SNR loss
Right	20	Severe
Left	22	Severe
Bilateral	17	Severe

Abbreviations: BKB-SIN, Bamford-Knowles-Bench Speech-in-Noise; CNC, consonant-nucleus-consonant; SNR, signal to noise ratio.

63.6 Additional Questions to the Reader

1. What does the information reported in Table 63.2 provide the clinician about this patient?
2. What advantages does bimodal listening offer for CI recipients?
3. How could the additional information gleaned from the MSTB be used even if the patient was not a CI candidate?
4. What is another important utility of speech recognition testing for CI patients other than determination of candidacy?

63.7 Discussion of Additional Questions to the Reader

1. What does the information reported in Table 63.2 provide the clinician about this patient?
   Use of the revised MSTB offers useful insights into this patient's communication abilities. First, although she continues to perform better on tests that provide more context (36%–54% on AzBio sentence materials as compared to 0%–20% on CNC Words), a more rapid rate of speech and multiple male and female talkers are more challenging (AzBio sentences as compared to HINT sentences where she understood 64%–88%). background noise presents a significant problem, although using bilateral amplification somewhat improves that challenging situation (i.e., signal to noise ratio [SNR] loss improving to 17 dB from 22 dB for the left-aided condition). The patient's ability to understand speech in background noise was evaluated in two ways using the MSTB. AzBio sentences are presented with the speech signal fixed at 60 dBA and multitalker babble presented at a +10 dB (easier) and +5 dB (more difficult) SNR. This measure allows the clinician to assess word recognition ability when noise is presented at two fixed SNRs. BKB-SIN also assesses speech in noise but is a pseudoadaptive test measure in which the SNR is gradually changing while the speech is consistently presented at 60 dBA. This measure assesses the SNR at which the patient can recognize 50% of words in the sentences. Both measures provide important insights into the individual's ability to successfully understand speech in noise. Finally, when context is largely removed (i.e., moving from sentences to individual words), this patient's reduced speech recognition is highlighted (i.e., decreasing from 36 to 54% for AzBio sentences to 0 to 20% using CNC words) making her ability to use supplementary information to understand speech more impressive.
2. What advantages does bimodal listening offer for CI recipients?
   Research suggests that there are many benefits to bimodal hearing (a hearing aid in one ear and a CI in the opposite ear) that bilateral conventional amplification cannot provide for individuals perceiving limited benefit with hearing aids. Research has demonstrated that bimodal recipients may achieve better scores in the bimodal condition on sentence measures, in quiet and noise, compared with scores in the preimplant everyday listening condition or with either a CI or a hearing aid alone. The combination of residual low-frequency acoustic hearing from hearing aids with greater fidelity with electric stimulation (i.e., CI) results in additional phonetic information that neither device alone can provide. This bimodal combination increases speech recognition, which is especially useful for challenging listening situations such as recognizing speech in background noise and appreciating music.
3. How could the additional information gleaned from the MSTB be used even if the patient was not a CI candidate?
   Patients typically perform best on tests involving open-set sentences presented in quiet. This is the test condition used in the United States for determining CI candidacy. It is not uncommon for patients to perform well under this condition and not meet candidacy criteria and still report significant difficulty communicating in their daily lives. Administration of the revised MSTB allows the clinician additional insight into the patient's abilities in a variety of listening conditions. This information should provide additional counseling regarding bilateral versus monaural hearing aid fitting, use of a frequency-modulated (FM) system or other assistive listening devices, and the patient's ability to use context and other communication strategies as supplements to auditory input. When coupled with verification of the patient's hearing aids, the clinician can also offer objective evidence of the potential need for more advanced technology and can counsel on specific features or accessories that may enhance the patient's listening challenges.
4. What is another important utility of speech recognition testing for CI patients other than determination of candidacy?
   Another important utility of speech recognition testing for CI recipients is longitudinal assessment of performance. Changes in performance can be tracked over time and used diagnostically to adjust programming parameters, troubleshoot equipment, or identify earlier decreases in performance that may be related to device failure.

63.8 Diagnosis and Recommended Treatment

Based on speech recognition ability reported in Table 63.2, in the best aided condition on open-set sentences in quiet using the MSTB, the patient was approved by the Hearing Implant Program Team for a CI in the right ear. Further, the team recommended bimodal listening for optimal speech recognition, particularly in challenging listening environments such as background noise. Typically, the team would have recommended a CI in the incrementally poorer left ear to allow for optimal bimodal listening; however, the patient had a history of left facial paralysis that raised concerns for recurrent facial paralysis following cochlear implantation. Thus the right ear was selected for the CI.

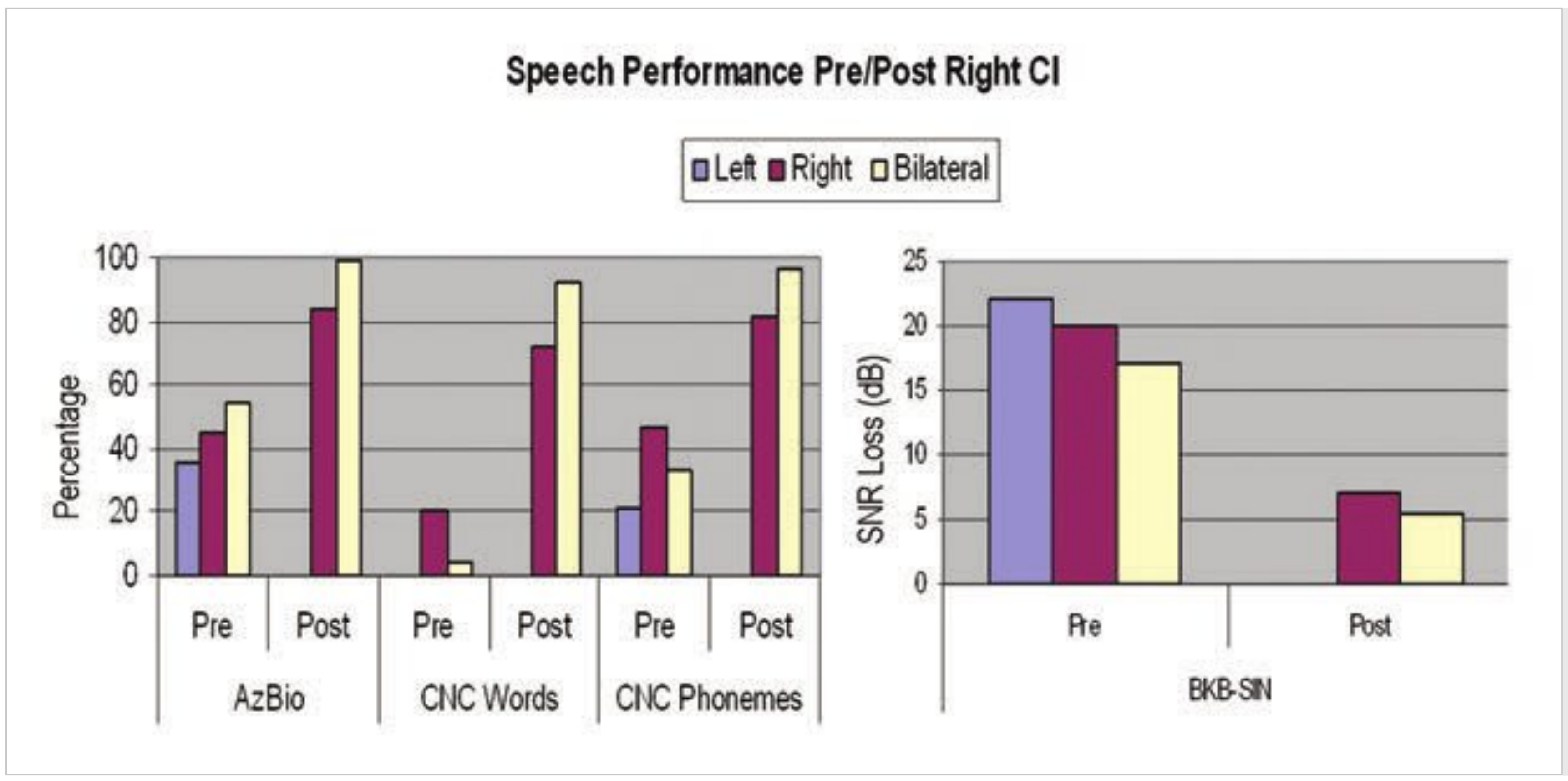


Fig. 63.2 Comparison of pre- and postoperative aided speech recognition scores.

63.9 Outcome

Following cochlear implantation, the patient's speech recognition score on sentence materials in quiet nearly doubled (compare □Table 63.2 to □Table 63.3 and illustrated in □Fig. 63.2 as pre and post scores). In addition, her speech recognition in noise significantly improved (compare □Table 63.2 to □Table 63.3 and illustrated in □Fig. 63.2 as pre and post scores) in the bimodal condition. The patient continues to use her FM system in the bimodal condition in complex listening environments.

Table 63.3 Postoperative speech recognition scores

Aided CNC words		
Test condition	Phonemes	Words
Right	79%	60%
Left	64%	36%
Bimodal	96%	88%
Aided AzBio (quiet)		
Test condition	Score	
Right	97%	
Left	42%	
Bimodal	96%	
Aided AzBio (+10 SNR)		
Test condition	Score	
Bimodal	92%	
Aided AZ BIO (+5 SNR)		
Test condition	Score	
Right	21%	
Bimodal	57%	
Aided BKB-SIN		
Test condition	Score	Degree
Right	12.5	Moderate
Bimodal	5.5	Mild

Abbreviations: BKB-SIN, Bamford-Knowles-Bench Speech-in-Noise; CNC, consonant-nucleus-consonant; SNR, signal to noise ratio.

63.10 Key Points

1. The use of various test measures can significantly alter the outcomes and interpretation for determination of candidacy for a CI.
2. It is important to use ecologically valid test measures that assess as many aspects of the patient's listening abilities as possible. This includes listening to sentences in quiet, lower-context words in quiet, and speech in noise.
3. Appropriate speech recognition materials can offer important information for determination of CI candidacy, recommendation of assistive technologies and hearing aid selection and programming, longitudinal assessment of performance, and counseling.

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64 Novel Use of Speech Recognition Testing for Counseling Cochlear Implant Recipients

Sarah A. Sydlowski

An adult cochlear implant (CI) recipient returned to a clinic reporting decreased performance in noisy listening environments with his bimodal fitting.

64.1 Clinical History and Description

AD is a 76-year-old male with a CI on one ear and a conventional hearing aid on the opposite ear. He had an appointment six months postimplantation of a Cochlear$^{\text{TM}}$ Americas (Centennial, CO) Nucleus Freedom Contour Advance (CI24RE-CA) electrode array and activation of his CP810 sound processor. At this visit, AD reported frustration when listening in background noise and also reported that he performs more poorly on speech recognition with the CI than he did with his bilateral hearing aids despite extensive auditory training after the implantation. AD had a history of progressive postlingual hearing loss for approximately 40 years that is likely related to occupational noise exposure. A preoperative audiological examination revealed a bilateral severe sensorineural hearing loss with word recognition scores (WRSs) of approximately 40%

AD wore appropriately fit behind-the-ear (BTE) hearing aids consistently since his hearing loss was initially diagnosed. Preoperative aided testing was conducted using AD's appropriately fit bilateral Phonak Naida S V SP BTE hearing aids (Phonak, U.S., Warrenville, IL). Testing was conducted with speech and noise, when applicable, presented from a loudspeaker at 0° azimuth with an input level of 60 dBA in the right hearing aid only, left hearing aid only, and bilaterally aided conditions. The CI candidacy evaluation consisted of sentences presented in quiet (AzBio sentences), words presented in quiet (consonant-nucleus-consonant [CNC] words), and sentences presented with pseudoadaptive noise (Bamford-Knowles-Bench Speech in Noise test [BKB-SIN]) AzBio sentences consist of sentences presented in quiet by two male and two female speakers. BKB-SIN is a pseudoadaptive test measure in which the signal to noise ratio (SNR) is measured by gradually decreasing the level of the noise while keeping the level of the speech constant at 60 dBA. This measure assesses the SNR at which the patient can recognize 50% of the words in the sentences. His preoperative aided speech recognition ability is summarized in Table 64.1. Results suggest that AD had significant difficulty with speech recognition even when sentences and words were presented in quiet with the signal presented at a level of conversational speech. With these results, AD met the U.S. Food and Drug Administration (FDA) criteria for cochlear implantation in either ear and was subsequently implanted in the left ear. AD continued to use his Phonak Naida S V SP hearing aid in the right ear.

AD's history of progressive hearing loss and consistent hearing aid use combined with his residual preoperative hearing were positive factors that would suggest the likelihood of substantial benefit from cochlear implantation, particularly with continued use of his Phonak Naida S V SP hearing aid in the nonimplanted right ear. Further, AD has been receiving aural rehabilitation (AR) services weekly with a speech-language pathologist (SLP) and was highly motivated to improve his hearing. Recently, his wife reported that he seems to frequently miss what she is saying, and it almost seems as if AD is not paying attention. AD and his wife confirmed that AD is performing more poorly than he was in the weeks initially following activation. Further, there have been no other changes in AD's medical status.

64.2 Audiological Testing

Discussion with AD regarding his listening habits revealed that AD is wearing the CI and hearing aid all his waking hours. AD admitted to frequently adjusting various settings on his remote control, particularly when he was listening in background noise. AD had several programs consisting of identical MAPs with various SmartSound$^{\text{TM}}$ strategies applied. AD most frequently used his "Everyday" program (Adaptive Dynamic Range Optimization [ADRO] + Auto-Sensitivity Control [ASC]), but often switched to the "Noise" program (ADRO + ASC + Zoom) or the "Focus" program (ADRO + ASC + Beam) when in more challenging listening environments. Inspection of AD's remote control revealed the volume control was operating at a setting of 8 (out of an upper limit of 9) and the sensitivity setting was at a setting of 7 (out of an upper limit of 20). The volume control acts to increase the perceived loudness of sound by increasing the C levels (upper stimulation levels) within the MAP. The default volume setting is typically 5 or 6. The sensitivity setting controls the sensitivity of the microphone. Increasing or decreasing the sensitivity setting effectively shifts the audible input intensity range. The default sensitivity setting is typically

Table 64.1 Preoperative aided speech recognition scores

CNC words		
Test condition	Phonemes	Words
Right ear	64%	28%
Left ear	51%	16%
Bilateral	43%	24%
AzBio sentences		
Test condition	In quiet	
Right ear	19%	
Left ear	4%	
Bilateral	36%	
BKB-SIN		
Test condition	SNR loss	Degree
Right ear	12	Moderate
Left ear	15	Severe
Bilateral	13	Moderate

Abbreviations: BKB-SIN Bamford-Knowles-Bench Speech-in-Noise; CNC, consonant-nucleus-consonant; SNR, signal to noise ratio.

Table 64.2 Speech recognition results in quiet and noise using AD's settings

AzBio sentences		
Test condition	In quiet (AD's settings)	
CI only	30%	
Bimodal	42%	
BKB-SIN		
Test condition	SNR loss (AD's settings)	Degree SNR loss (AD settings)
Bimodal "Everyday" program with speech and noise at 0°	11	Moderate
Bimodal "Noise" program with speech at 0° noise at 180°	8.5	Moderate
Bimodal "Focus" program with speech at 0° noise at 180°	5.0	Mild

Abbreviations: CI, cochlear implant; SNR, signal to noise ratio.

10 or 12 for quiet listening. AD reported that the settings he is currently using have been typical for him. Testing using speech in quiet (AzBio sentences) and noise (BKB-SIN) was conducted in a double-walled, sound-treated room using an input level of 60 dBA with the patient using the setting described above (Table 64.2).

64.3 Questions to the Reader

1. What do the results in Table 64.2 indicate?
2. What are the SmartSound™ strategies used by AD and how do the strategies work?
3. How do the volume control and sensitivity settings operate in the Cochlear™ Americas CI system?

64.4 Discussion of Questions to the Reader

1. What do the results in Table 64.2 indicate?
   Comparison of longitudinal speech recognition scores can provide important information regarding changes in performance over time or in different listening conditions. Determination of clinical significance of any changes can be determined by referring to published confidence intervals for the test of interest. In this case, speech recognition testing (Table 64.2) revealed a clinically significant improvement for AzBio sentence recognition in quiet using the CI alone versus the hearing aid (30% compared with 4%); however, AD's CI scores are still lower than anticipated 6 months post-activation. Given his history of residual hearing and appropriate hearing aid use, a clinician would expect speech recognition of sentences to be at least 50 to 60%, if not greater. Additionally, there was no clinically significant improvement between the bilateral hearing aid (36%) (Table 64.1) and bimodal (CI + HA) (42%) (Table 64.2) conditions. This finding is consistent with AD's report that he does not perceive his hearing has improved using his CI. In fact, AD's performance for sentences in quiet in the bimodal condition (Table 64.2) is essentially equal to his preoperative performance with bilateral hearing aids (Table 64.1). Similarly, speech in noise (BKB-SIN) testing in the "Everyday" program revealed no significant difference between his preoperative (Table 64.1) and postoperative scores (Table 64.2). AD demonstrated better speech in noise recognition with his "Noise" and "Focus" programs with noise directed at 180° (Table 64.2), suggesting these programs may be beneficial if AD can position himself with background noise behind him and the speaker to his front. AD, however, is still exhibiting mild to moderate difficulty in background noise (Table 64.2).
2. What are the SmartSound™ strategies used by AD and how do the strategies work?
   SmartSound™ describes preprocessing strategies that are applied to an incoming acoustic signal to optimize the recipient's listening abilities in a variety of listening situations. The inclusion of ADRO optimizes the incoming signal for improved speech recognition. Specifically, ADRO increases the audibility of soft sounds, maintains a comfort level for higher input sounds, and attempts to position the input speech signal to lie within the recipient's electric dynamic range by manipulating channel gains. ASC, as previously described, tries to maintain the intensity of the speech signal above the intensity of the surrounding background noise. Once the ASC breakpoint is reached (default = 57 dB SPL), ASC is activated. Both ADRO and ASC were active in the "Everyday," "Noise," and "Focus" programs AD was using. Additionally, the "Noise" and "Focus" programs use preprocessing strategies combined with ADRO and ASC to optimize speech recognition in background noise, particularly if the noise is directed to the back of the CI recipient with speech to the front. The "Noise" program uses the Zoom algorithm, which is a fixed directional microphone with a null at approximately 120° azimuth. The goal of the "Noise" program is to improve the audibility of speech in background noise when there is a talker located directly in front of the CI recipient. Finally, "Focus" utilizes the Beam algorithm, which is an adaptive directional microphone that attenuates noise from the sides and back of the CI recipient by changing the position of the nulls within the polar design of the directional microphone. This reportedly provides improved audibility and recognition of talkers from the front.
3. How do volume control and sensitivity settings operate in the Cochlear™ Americas CI system?
   The volume control and sensitivity settings are useful tools available to CI recipients allowing for modification and adjustment of the acoustic input signal. The volume control setting is used by the CI recipient to adjust comfort levels up or down. The sensitivity setting controls the audibility of soft sounds and the compression of loud sounds. CI recipients may increase the sensitivity setting to improve the audibility of soft or distant sounds and may decrease the sensitivity setting if environmental sounds are too intense. When these adjustments are made, instantaneous input dynamic range

(IIDR) remains constant. Because the IIDR remains constant, increasing the sensitivity setting to capture softer sounds will also decrease the intensity of sounds that will be compressed. Conversely, decreasing the sensitivity setting to reduce softer input levels will also allow higher input levels to be processed without compression. The ASC feature theoretically eliminates the need for manual control of sensitivity by changing the sensitivity setting automatically, depending on the listening environment. The user-selected ASC setting (default = 12) defines the upper limit.

64.5 Additional Testing

Speech recognition testing was repeated in the same conditions as reported in Table 64.2 using the recommended volume control (5) and sensitivity (12) settings (Table 64.3) to allow for comparison of AD's performance using his current preferred settings and the recommended settings. These test conditions included sentences in quiet (AzBio sentences) and sentences in noise (BKB-SIN). For BKB-SIN, noise was presented at 0° when using the "Everyday" program and at 180° when using the "Noise" and "Focus" programs.

64.6 Additional Questions to the Reader

1. How do these results (Table 64.3) compare with the results in Table 64.2 using AD's preferred settings?
2. Based on the results of testing with the recommended default settings (Table 64.3), how would a clinician counsel AD? Do these results suggest the possibility of significant improvement?
3. What information obtained after changing the volume control and sensitivity settings would be most relevant for developing a clinical action plan?
4. What else should the clinician consider investigating when addressing reported decrease in speech recognition in a CI recipient?

64.7 Discussion of Additional Questions to the Reader

1. How do these results (Table 64.3) compare with the results in Table 64.2 using AD's preferred settings?
   Results reported in Table 64.3 indicated clinically significant improvement on all measures when the volume control and sensitivity settings were adjusted appropriately. Sentence recognition improved to levels that would be more typical of a CI recipient with AD's hearing and aural rehabilitation history. Additionally, AD demonstrated a clinically significant improvement in the bimodal condition as compared with the CI-only condition as would be expected. Speech-in-noise ability in the bimodal condition improved such that AD demonstrated only a mild difficulty in background noise.
2. Based on the results of testing with recommended default settings (Table 64.3), how would a clinician counsel AD? Do these results suggest the possibility of significant improvement?
   Results reported in Table 64.3 demonstrated clinically significant improvement in using all three programs with the clinic-recommended volume control and sensitivity settings when compared with AD's current user settings. For example, the SNR necessary for AD to recognize 50%of the target words improved by 5 dB. One may question the significance of an improvement of 5 dB SNR; however, Nilsson et al demonstrated that every 1 dB SNR improvement equates to an 8.5%improvement in speech recognition when using Hearing in Noise Test (HINT). This finding highlights how small improvements in SNR loss can greatly increase the ability of a patient to recognize speech in background noise. AD was counseled concerning the potential negative impact of injudicious adjustment of controls and how these changes can result in frustration and reduced speech recognition.
3. What information obtained after changing the volume control and sensitivity settings would be most relevant for developing a clinical action plan?
   Obtaining a detailed case history including current performance compared with preoperative and prior CI performance,

Table 64.3 Comparison of speech recognition in quiet and noise using AD's settings (volume control at 8, sensitivity setting at 7) and recommended settings (volume control at 5, sensitivity setting at 12)

AzBio sentences				
Test condition	In quiet(AD's settings)	In quiet(recommended settings)		
CI only	30%	72%		
Bimodal	42%	88%		
BKB-SIN				
Test condition	SNR loss(AD's settings)	Degree SNR loss (AD's settings)	SNR loss(recommended settings)	Degree SNR loss (recommended settings)
Bimodal "Everyday" program with speech and noise at 0°	11 dB	Moderate	6.5 dB	Mild
Bimodal "Noise" program with speech at 0° and noise 180°	8.5 dB	Moderate	3.5 dB	Mild
Bimodal "Focus" program with speech at 0° and noise at 180°	5.0 dB	Mild	0 dB	Normal/near-normal

Abbreviations: BKB-SIN, Bamford-Knowles-Bench Speech-in-Noise; CI, cochlear implant; SNR, signal to noise ratio.

challenging situations, and habits regarding program adjustment were instrumental in developing initial suspicions of AD's program/remote control misuse. Although speech recognition testing is often only completed with the speech processor at user settings, modifying the protocol to objectively compare AD's current volume control and sensitivity settings with recommended settings provided evidence for AD that his adjustments were negatively impacting his hearing ability. Although developing a standard protocol for most patients is important for consistency of care, the question of interest may necessitate considering alternative methods of objectively identifying the current patient problem. Critical clinical analysis and modification of clinical protocols can be extremely helpful in the efficient and accurate identification of the source of the problem as well as proving to be useful for effective counseling.

4. What else should the clinician consider investigating when addressing reported decrease in speech recognition in a CI recipient?
   When comparing results of longitudinal speech recognition testing, the test condition is important to consider. Changes to speech recognition in the bimodal condition may reflect changes in hearing levels and related hearing aid adjustments, changes to the CI side, or both the CI and hearing aid. Focusing on the speech recognition results when testing in the CI-alone condition, other possible causes of a decrease in speech recognition could include the need for reprogramming, external equipment disrepair such as a faulty cable/coil or clogged microphone filter, or changes in medical status such as cognitive decline or stroke. Decreases in speech recognition without resolution following reprogramming that are otherwise unexplained could be an early sign of device failure.

64.8 Diagnosis and Recommended Treatment

AD's CI is functioning well. AD's hearing aid is programmed appropriately and his hearing loss has not changed. AD's reported decline in speech recognition in noise was related to his inappropriate use of the recommended setting on the CI. Based on results of testing, no programming changes were made other than to lock the sensitivity control at 12 (with the patient's consent) and decrease the sensitivity breakpoint to 50 dB SPL to allow the ASC to activate at lower input levels of background noise. AD was also counseled extensively about appropriate use (and the consequences of misuse) of his settings and positioning himself optimally for conversations in background noise.

64.9 Outcome

AD returned 2 weeks later and reported his marriage had been saved and he could not believe how much better he was hearing. AD is making judicious changes to the volume control setting as needed and is selecting the appropriate programs based on his listening environment.

64.10 Key Points

1. This case report highlights the value of using a thorough case history and objective verification in a sound-treated room to evaluate the cause of the patient's report rather than focusing primarily on programming adjustments alone.
2. It is important to approach each patient as a unique individual and to modify clinical protocols to address specific patient reports.
3. This case demonstrates the importance of counseling patients and empowering patients to understand their hearing loss and how best to manage their hearing loss though appropriate use of their hearing devices.

Suggested Reading

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Part 6

Hearing Assistive Technology

65 FM Technology Improves Quality of Life 284

66 Recommendation and Verification of a Personally Worn FM System 288

65 FM Technology Improves Quality of Life

Michelle Arnold, Theresa Chisolm, Paula J. Myers, and Gabrielle H. Saunders

This case report discusses management of a patient with auditory processing difficulties following multiple blast exposures and probable mild traumatic brain injury (mTBI) during deployment while serving in the United States Marine Corps.

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65.1 Clinical History and Description

RJ is a 23-year-old male Marine Corp veteran who completed one tour of duty during Operation Iraqi Freedom (OIF) in 2008 and one tour of duty during Operation Enduring Freedom (OEF) in 2010 to 2011. During both tours, RJ was stationed in combat scenarios and sustained approximately five blast injuries while on various missions. RJ reported the most damaging blast exposure occurred while he was on foot. The close-range improvised explosive device (IED) emitted a blast wind to his right side that nearly knocked him off his feet. RJ stated that he immediately experienced disorientation, confusion, a dazed feeling, right ear tinnitus, and headache. Additionally, RJ reported being involved in two vehicle accidents resulting from IED blasts during his 2008 deployment. Although RJ was seated in an armored vehicle on both occasions, the blasts resulted in his feeling dazed and confused with headache. RJ did not use hearing protection while deployed. RJ also reported that during this second tour of duty he began noticing hearing and memory difficulties.

Following his honorable discharge, RJ used his GI Bill benefits to attend college as a first-time undergraduate student. He experienced no postconcussive symptoms prior to his reported blast exposures that occurred during combat. Recently, RJ scheduled an appointment at a Veterans Administration (VA) audiology clinic with symptoms of hearing loss, constant right ear tinnitus, difficulty hearing in quiet and noise, dizziness/imbalance, memory loss, difficulty concentrating in class, and problems sleeping.

65.2 Audiological Testing

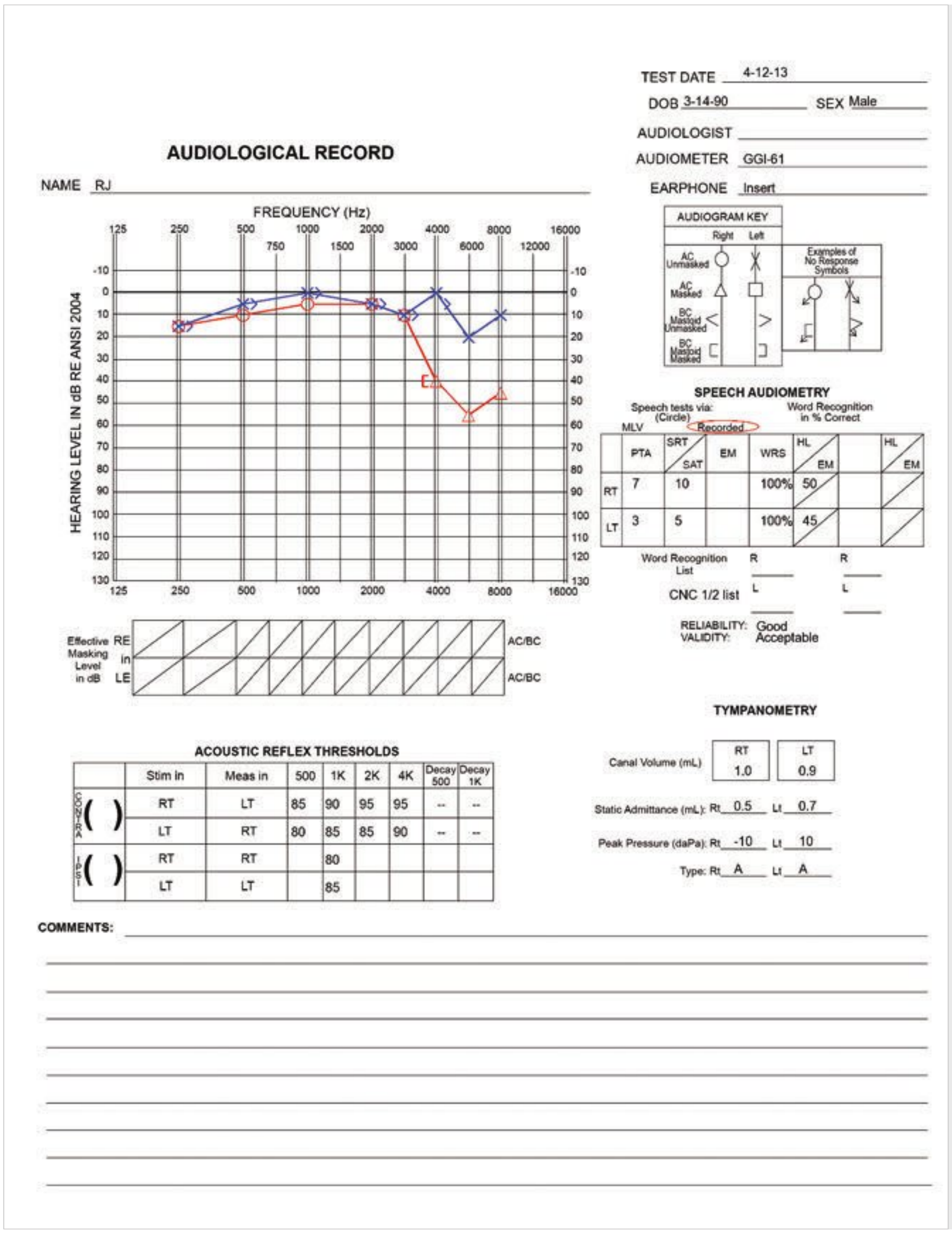
Conventional audiological examination included immittance, distortion-product otoacoustic emissions (DPOAEs), pure-tone and speech audiometry. Pure-tone audiometry revealed normal hearing in the right ear at 250 to 3000 Hz followed by a mild to moderate high-frequency sensorineural hearing loss (HFSNHL) at 4000 to 8000 Hz. Pure-tone thresholds for the left ear revealed normal hearing at 250 to 8000 Hz with the exception of a slight hearing loss at 6000 Hz. (Fig. 65.1). Speech audiometry revealed that RJ's speech recognition thresholds (SRTs) were in agreement with pure-tone average and normal ability to receive speech bilaterally. Word recognition scores (WRSs) indicated normal ability to recognize speech bilaterally. Immittance testing resulted in bilateral type A tympanograms with normal ear canal volume (mL), static admittance (mL), and middle ear pressure (daPa). Bilateral ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present at expected levels at 500 to 4000 Hz. Acoustic reflex decay was negative at 500 Hz and 1000 Hz bilaterally. Results of RJ's audiological evaluation prompted referrals for further testing and treatment to address his functional communication concerns.

65.3 Questions to the Reader

1. What factors in RJ's history may be contributing to the functional hearing difficulties he reports?
2. What situations in RJ's daily listening environments might be most affected by RJ's hearing loss and functional hearing complaints?
3. What might help explain RJ's asymmetric HFSNHL and tinnitus?
4. What additional testing could be recommended for RJ?

65.4 Discussion of Questions to the Reader

1. What factors in RJ's history may be contributing to the functional hearing difficulties he reports?
   It is possible that RJ's memory and concentration difficulties might be a manifestation of posttraumatic stress disorder (PTSD) due to the extensive exposure to combat he endured throughout his military deployments. Many veterans who are exposed to heavy combat are diagnosed with PTSD following discharge from the military. PTSD symptoms can mimic postconcussive symptoms. In fact, RJ was evaluated for and diagnosed with PTSD at this VA medical center shortly after his audiological evaluation. Another contributing factor might be the HFSNHL in RJ's right ear. Although the right ear HFSNHL is mild to moderate, and his left ear hearing is still within normal limits, RJ may be experiencing difficulties localizing sound and understanding high-frequency speech information when presented to his right side.
2. What situations in RJ's daily listening environments might be most affected by RJ's hearing loss and functional hearing complaints?
   During RJ's initial appointment, he reported that he was a first-time college student under the GI Bill. RJ stated he was experiencing difficulties during college lectures, particularly in larger lecture halls. When questioned further by the clinician, it was revealed that RJ was easily distracted by the whispers and movements of other students around him during classes, and found the reverberation in large lecture halls especially problematic. In fact, RJ reported that sometimes it was "nearly impossible" to understand and attend to lectures. This could be a result of his mTBI, his right ear HFSNHL, or both. Another daily listening environment affected by his



AUDIOLOGICAL RECORD

NAME RJ

TEST DATE 4-12-13

DOB 3-14-90 SEX Male

AUDIOLOGIST

AUDIOMETER GGI-61

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	7	10		100%	50		
LT	3	5		100%	45		

Word Recognition List R L R L

CNC 1/2 list

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL)	RT	LT
	1.0	0.9

Static Admittance (mL): Rt 0.5 Lt 0.7

Peak Pressure (daPa): Rt -10 Lt 10

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	85	90	95	95	--	--
	LT	RT	80	85	85	90	--	--
IPSI ( )	RT	RT		80				
	LT	LT		85				

COMMENTS:

Fig. 65.1 Audiometric results.

blast exposure and subsequent hearing loss involved RJ's wife and their mutual friends. RJ reported that he has difficulties communicating with his wife and friends when in noisy restaurants and bars, which they frequent regularly. RJ reported that evenings out with his wife and friends were fatiguing and frustrating because it was so difficult to recognize speech, particularly in noise, and because he had to concentrate very carefully in order to engage in conversation. Further, RJ expressed feelings of guilt and anxiety when discussing the communication barriers he was experiencing with his wife, noting that he was worried she thought he was not paying attention.

3. What might help explain RJ's asymmetric HFSNHL and tinnitus?
   RJ's asymmetric right ear HFSNHL and constant tinnitus were likely caused by the right ear's close proximity to the IED blast he described during his initial appointment. Note that RJ did not report the use of any hearing protection for any of the blast exposures he sustained.
4. What additional testing could be recommended for RJ?
   The functional listening difficulties RJ described during his initial appointment coupled with his close-range blast exposure prompted the clinician to refer RJ for an evaluation of his auditory processing abilities. In addition to assessment of RJ's functional hearing abilities and auditory processing abilities, RJ was referred for a comprehensive vestibular evaluation that included ocular motor testing, positional testing, bithermal caloric testing, and rotational chair evaluation to assess his reported dizziness. All vestibular tests were within normal limits with the exception of a left peripheral weakness that was revealed by rotational chair evaluation and left-side water calorics. RJ was subsequently diagnosed with left benign paroxysmal positional vertigo (BPPV). BPPV can occur as a result of mTBI if the head injury causes calcium crystals (otoconia) in the utricle to become dislodged. Oftentimes, BPPV rectifies itself over time as the otoconia are absorbed into the endolymph of the semicircular canal; however, RJ's BPPV was effectively treated with canalith repositioning by a vestibular specialist. RJ was also referred to Group Progressive Tinnitus Management sessions to address his report of tinnitus, but RJ declined, stating his tinnitus was not particularly bothersome. RJ was counseled on the importance of hearing protection when in excessive noise. Although his right ear asymmetry and tinnitus were likely

caused by the close-range IED blast, RJ was additionally referred to otolaryngology for evaluation to rule out right-side retrocochlear pathology. Magnetic resonance imaging (MRI) testing of the right temporal bone with gadolinium contrast revealed no abnormalities.

65.5 Additional Testing: Assessment of Functional Hearing Ability

Tests examining RJ's functional hearing ability and communication concerns, as well as tests of auditory processing, were administered at a follow-up visit with his VA audiologist. Testing included the Performance Perceptual Test (PPT), the Adaptive Test of Temporal Resolution (ATTR), a time compressed sentences test (TCST), and the Staggered Spondaic Word (SSW) test.

The PPT is a test of speech in noise that assesses both perceived and actual ability to understand speech in noise by using the same test materials (the hearing in noise test [HINT] sentences), the same unit of measurement (dB signal to noise [S/N]) for both comparisons, and an adaptive testing format. By comparing measured and perceived ability, a direct measure of the extent to which individuals misjudge their hearing ability is obtained. To assess actual ability, the signal to noise ratio (SNR) for 50% correct performance is determined. In the sound field the patient listens to the HINT sentences presented at 65 dB sound pressure level (SPL) in noise and repeats back as much or as little of each sentence as possible. If all keywords in a sentence are repeated correctly, the SNR is made more adverse. If one or more keywords are repeated incorrectly, the SNR is made less adverse. To assess perceived ability, the SNR at which patients perceive they can "just understand everything" is obtained. Once again, patients listen to HINT sentences in the sound field. After hearing each sentence, the patient says either, "Yes, I could understand everything," or, "No, I could not understand everything." If the patient answers "yes," the SNR is made more adverse; if the patient answers "no," the SNR is made less adverse. By subtracting the perceived SNR from the performance SNR, the performance-perceptual discrepancy (PPDIS) is obtained. The PPDIS is a measure of the accuracy to which individuals (mis)judge their hearing ability in noise. A negative PPDIS implies the participant selected a less adverse SNR than that at which he or she can actually perform (i.e., the listener underestimates his or her own hearing ability). A positive PPDIS implies the participant selected a more adverse SNR than that at which he or she can perform (i.e., the listener overestimates his or her own hearing ability). RJ's actual binaural speech recognition as measured by the PPT was 13.7 dB, and his perceived binaural speech recognition was 14.0 dB. His PPDIS was therefore 0.7 dB.

The ATTR is an assessment of temporal resolution that measures binaural gap detection (i.e., the smallest gap a patient is able to detect in noise). Gap detection tasks involve listening to two or more bursts of noise, one of which includes a silent interval embedded in the noise. The ATTR uses an adaptive format in which gaps as small as 1 ms are embedded in diotic noise centered around 1000 Hz. Using a two-alternative forced-choice paradigm, the patient is required to select the interval that contained the gap. If the patient selects incorrectly, the following interval contains a larger gap, and if the patient selects correctly, the following interval contains a smaller gap. Performance on measures of gap detection is thought to be related to processing of degraded speech, such as speech in noise. RJ's binaural gap detection threshold was 3.26 ms.

The TCST was developed to assess speech recognition for rapid speech. Sentences are presented in quiet at a normal rate and between 40 and 60%temporal compression without degrading the spectral information of the sentences. Patients repeat back each sentence after it is presented, and a key words percent correct score is obtained. This test was selected for RJ because processing deficits for rapid speech have been shown to be associated with blast injury. RJ was able to repeat all target words on the TCST at 50 and 60%compression with 100%accuracy.

The SSW test was developed by to assess binaural separation and the interhemispheric transfer of auditory information via the corpus collosum using a dichotic listening task. Two spondee words are presented at each trial, with the participant being asked to repeat the words in the exact presentation order. The spondees are presented in a "staggered" fashion (i.e., the last syllable of the first spondee is heard in one ear simultaneously with the first syllable of the second spondee in the opposite ear). This test was selected for RJ because it has been shown to be sensitive to processing deficiencies in veterans with blast exposure. RJ was able to repeat all spondaic words on the SSW with 100%accuracy.

65.6 Diagnosis and Recommended Treatment

RJ was diagnosed with right ear HFSNHL and subjective tinnitus and left BPPV. RJ revealed normal performance on all tests other than the PPT, on which his performance speech reception thresholds in noise (actual speech recognition) were poorer than the age-appropriate norms. RJ continued to report substantial functional listening difficulties despite normal peripheral hearing in the left ear and a mild HFSNHL in the right ear. To address his concerns, RJ was offered computerized auditory training (Brain Fitness Program, Posit Science Corp., San Francisco, CA) combined with provision of a personal FM system (Phonak iSense bilateral receivers and ZoomLink+transmitter, Phonak U.S., Warrenville, IL) (□ Fig. 65.2 a, b). The Brain Fitness Program (BFP) consists of six exercises designed to enhance

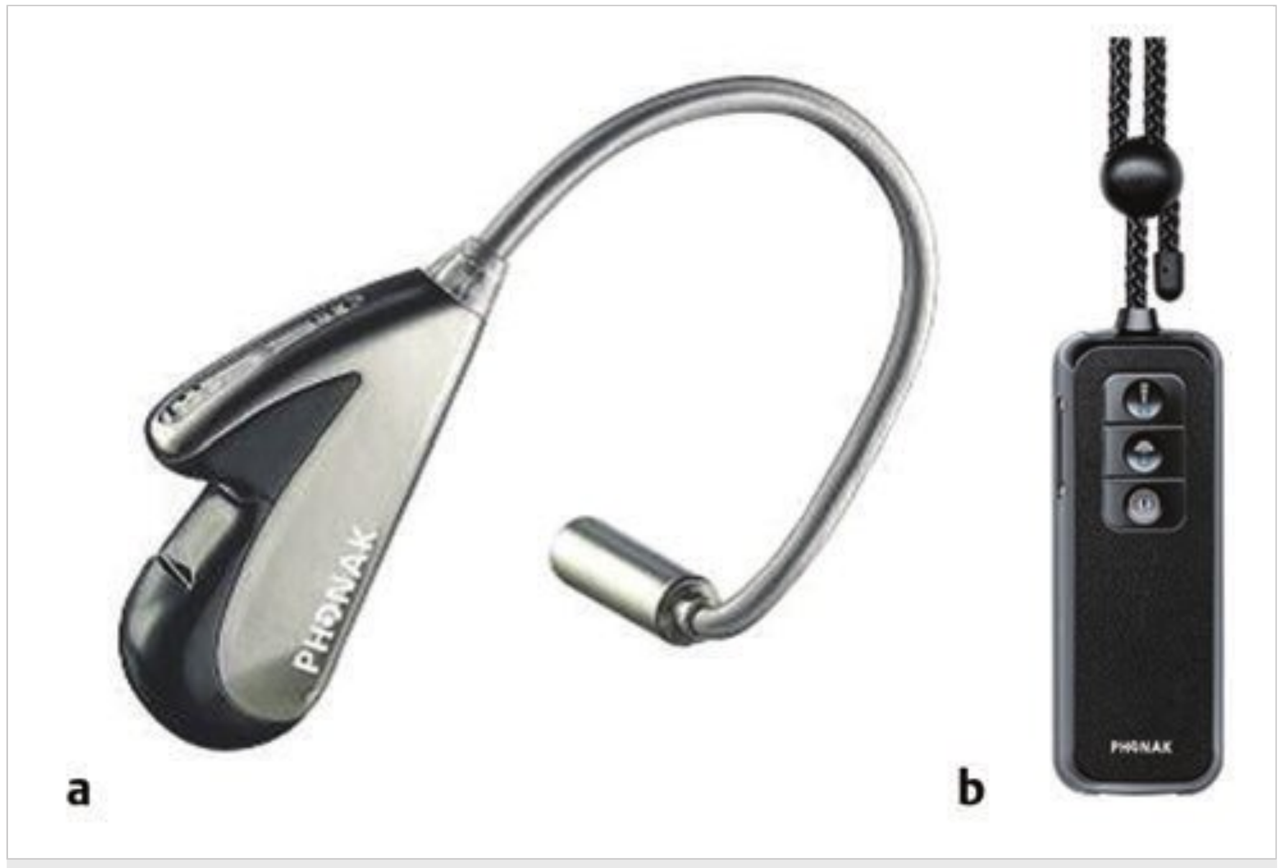


Fig. 65.2 Phonak iSense Micro (a) and ZoomLink+(b).

temporal processing and auditory working memory skills through intensive practice listening to temporally altered signals and changing memory loads. The exercises are adaptive, such that task difficulty automatically adjusts based on the user's performance. The reader should note that as of 2012, the BFP program is now incorporated into the Posit Science online BrainHQ program, which is available as a monthly subscription. For more information on Brain Fitness or Brain HQ, see http://positscience.com.

65.7 Outcome

RJ returned 1 month later for an appointment after receiving his treatment plan during which repeat testing on the PPT was performed and subjective outcome questionnaires were administered. Follow-up testing on the PPT revealed an SNR advantage of approximately + 14 when RJ used the FM system. This finding illustrates that the FM system enabled RJ to overcome his significant SNR loss revealed with the PPT. Improvements were also seen on the self-report outcome measures that RJ completed at his follow-up appointment. The majority of responses on a self-report questionnaire regarding auditory processing skills reflected self-perceived improvement in auditory memory, confidence performing auditory processing-related tasks, and processing speed. Likewise, 6 of 26 items on a generic quality of life questionnaire reflected self-perceived improvements.

RJ only completed 5 of the 20 recommended hours of auditory training with the BFP, but he reported using the FM system at least 3 hours a day. RJ said he found the Phonak iSense FM system particularly helpful in class, and it significantly improved his daily quality of life. His wife, who was encouraged to accompany him to his audiology appointments, also commented that the FM system "changed both of our lives for the better." RJ decided to retain his FM system at the end of the trial period.

65.8 Key Points

1. Many thousands of Operation Iraqi Freedom and Operation Enduring Freedom veterans are presenting in VA clinics nationwide with a wide array of auditory and vestibular deficits following tour(s) of duty. Many of these patients are also presenting to university and hospital audiology clinics and private practices. Conventional audiological examination on many of these patients reveals normal or near-normal peripheral hearing sensitivity, yet auditory complaints continue to persist. Patients with a history of mTBI who present with persistent functional auditory deficits despite normal audiological results should be referred for further assessment and treatment of their functional hearing and communication difficulties. Part of this assessment may include auditory processing testing.
2. Although most of the objective auditory processing test results did not reveal any obvious listening deficits for this patient, RJ's binaural performance SRTs in noise revealed below normal ability to recognize speech in noise. This deficit was reflected in his subjective reports of listening difficulties. Furthermore, RJ's listening needs in environments such as the classroom or social gatherings warranted an FM trial. Engaging in patient-centered care and addressing RJ's individual concerns regarding his academic performance and social life resulted in positive outcomes that were appreciated not only by RJ but by his wife as well.

Suggested Reading

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66 Recommendation and Verification of a Personally Worn FM System

Alison M. Brockmeyer-Lauer

GB, a 68-year-old female seen for audiological testing and treatment, is fit with bilateral hearing aids and a frequency-modulated (FM) system.

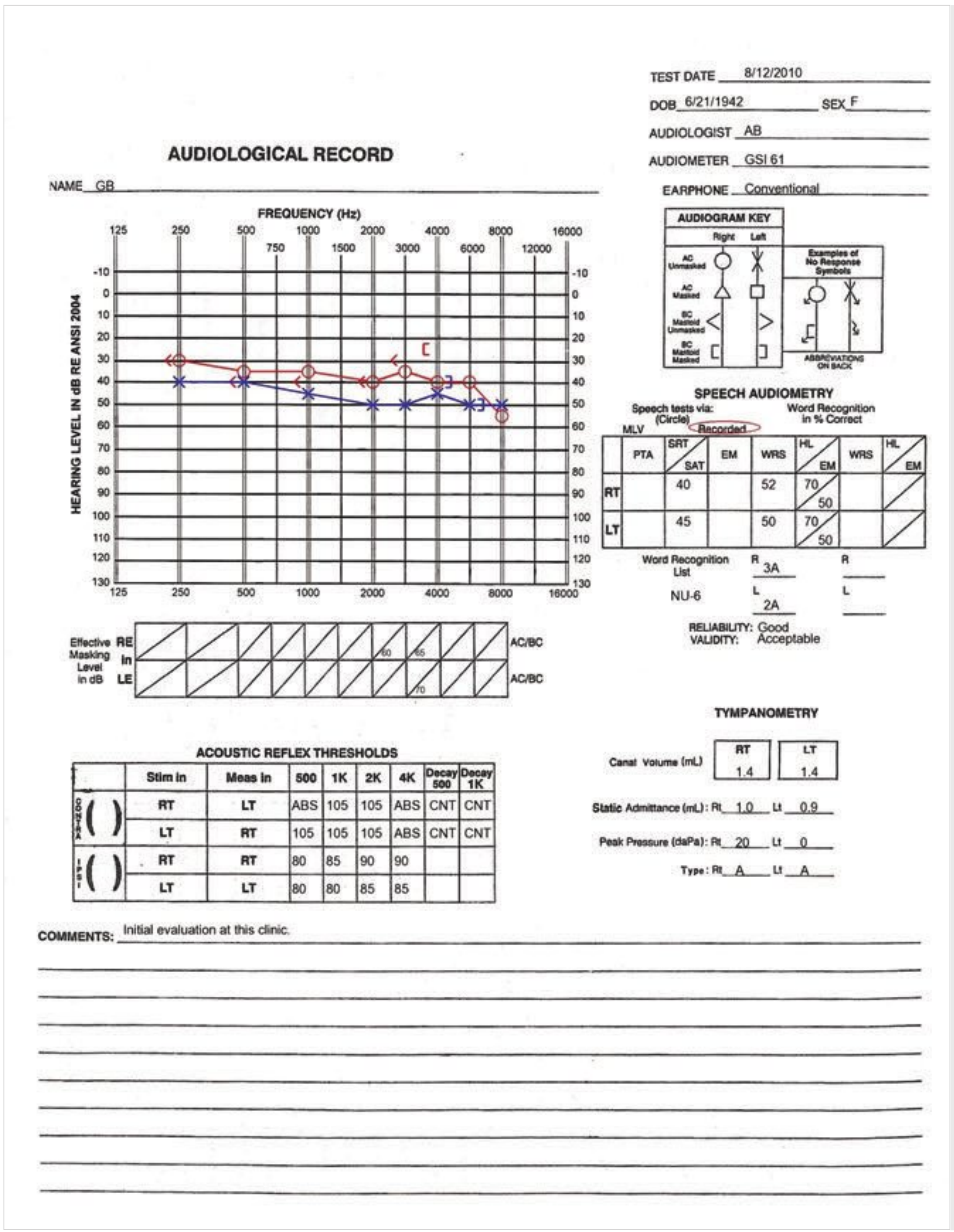
66.1 Clinical History and Description

GB is a 68-year-old female who presents with known bilateral hearing loss, which was first evident in high school. She reports constant tinnitus bilaterally, with the tinnitus louder in the left ear. She denies dizziness but reports that she experienced positional vertigo approximately 1 year ago. The dizziness, however, resolved after treatment with the Epley maneuver. She denies otalgia and excessive noise exposure but reports a familial history of progressive hearing loss in her father.

GB reports her hearing loss has progressed, and she began investigating hearing aids approximately 15 years ago. She is currently wearing bilateral behind-the-ear (BTE) hearing aids. She reports she does not hear well with her current hearing aids, especially in group settings and noisy environments. GB states she is constantly asking others to repeat even when wearing her hearing aids, and she uses captions on the television to follow the dialogue. GB reports she complained to her audiologist of continued hearing difficulty, and the audiologist increased the overall gain several times in her hearing aids. The increased gain, however, has not improved her communication, and she reports using her volume control often to reduce the loudness to prevent discomfort.

66.2 Audiological Testing

Audiological examination (Fig. 66.1) revealed mild sensorineural hearing loss from 250 to 6000 Hz, sloping to a moderate loss at 8000 Hz in the right ear. The left ear presented with mild



AUDIOLOGICAL RECORD

NAME GB

TEST DATE 8/12/2010

DOB 6/21/1942 SEX F

AUDIOLOGIST AB

AUDIOMETER GSI 61

EARPHONE Conventional

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM	WRS	HL / EM
RT		40		52	70 / 50		
LT		45		50	70 / 50		

Word Recognition List NU-6

R 3A R

L 2A L

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	1.4	1.4

Static Admittance (mL): Rt 1.0 Lt 0.9

Peak Pressure (daPa): Rt 20 Lt 0

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas In	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT	ABS	105	105	ABS	CNT	CNT
	LT	RT	105	105	105	ABS	CNT	CNT
IPSI ( )	RT	RT	80	85	90	90		
	LT	LT	80	80	85	85		

COMMENTS: Initial evaluation at this clinic.

Fig. 66.1 GB's initial audiological examination

Table 66.1 GB's frequency-specific LDLs in dB HL and dB SPL

Right ear		
Frequency	dB HL	dB SPL
500 Hz	80	94.4
1000 Hz	90	102.0
2000 Hz	80	95.1
3000 Hz	85	103.0
4000 Hz	85	104.0
Left ear		
Frequency	dB HL	dB SPL
500 Hz	80	95.5
1000 Hz	90	100.7
2000 Hz	85	103.2
3000 Hz	75	96.1
4000 Hz	85	96.1

Abbreviations: HL, hearing level; LDLs, loudness discomfort levels; SPL, sound pressure level.

sensorineural hearing loss from 250 to 500 Hz, sloping to a moderate sensorineural hearing loss from 1000 to 8000 Hz. The speech recognition threshold (SRT) was mildly impaired in the right ear and moderately impaired in the left ear. Word recognition scores (WRSs) were obtained using a recorded version of Northwestern University Auditory Test No. 6 (NU-6) monosyllabic word lists with a female speaker. Words were presented at most intelligible level (MIL), corresponding to a 40 dB sensation level (dB SL) relative to the SRT. Scores of 52% and 50% were obtained in the right and left ears, respectively, demonstrating poor word recognition ability bilaterally. Immittance audiometry was performed and tympanograms and ipsilateral acoustic reflex thresholds (ARTs) from 500 to 4000 Hz were within normal limits bilaterally. Contralateral ARTs were elevated or absent from 500 to 4000 Hz bilaterally. Contralateral acoustic reflex decay could not be performed due to the elevated or absent contralateral ARTs.

Frequency-specific loudness judgments were performed under headphones (Table 66.1). A probe tube from a probe microphone was placed in the ear canal under the headphones in order to directly measure loudness discomfort levels (LDLs) in dB sound pressure level (dB SPL). LDLs were measured bilaterally at 500 to 4000 Hz and the levels judged as "loud, but OK" were recorded in dB hearing level (HL) and dB SPL.

66.3 Questions to the Reader

1. How would you counsel the patient on the results of the audiological examination and specifically focusing on the WRSs?
2. What would you recommend to the patient to improve communication?
3. How can hearing assistive technology (HAT) improve the patient's communication compared with bilateral hearing aids alone?
4. What type of verification is required to fit an FM system?

66.4 Discussion of Questions to the Reader

1. How would you counsel the patient on the results of the audiological examination and specifically focusing on the WRSs?

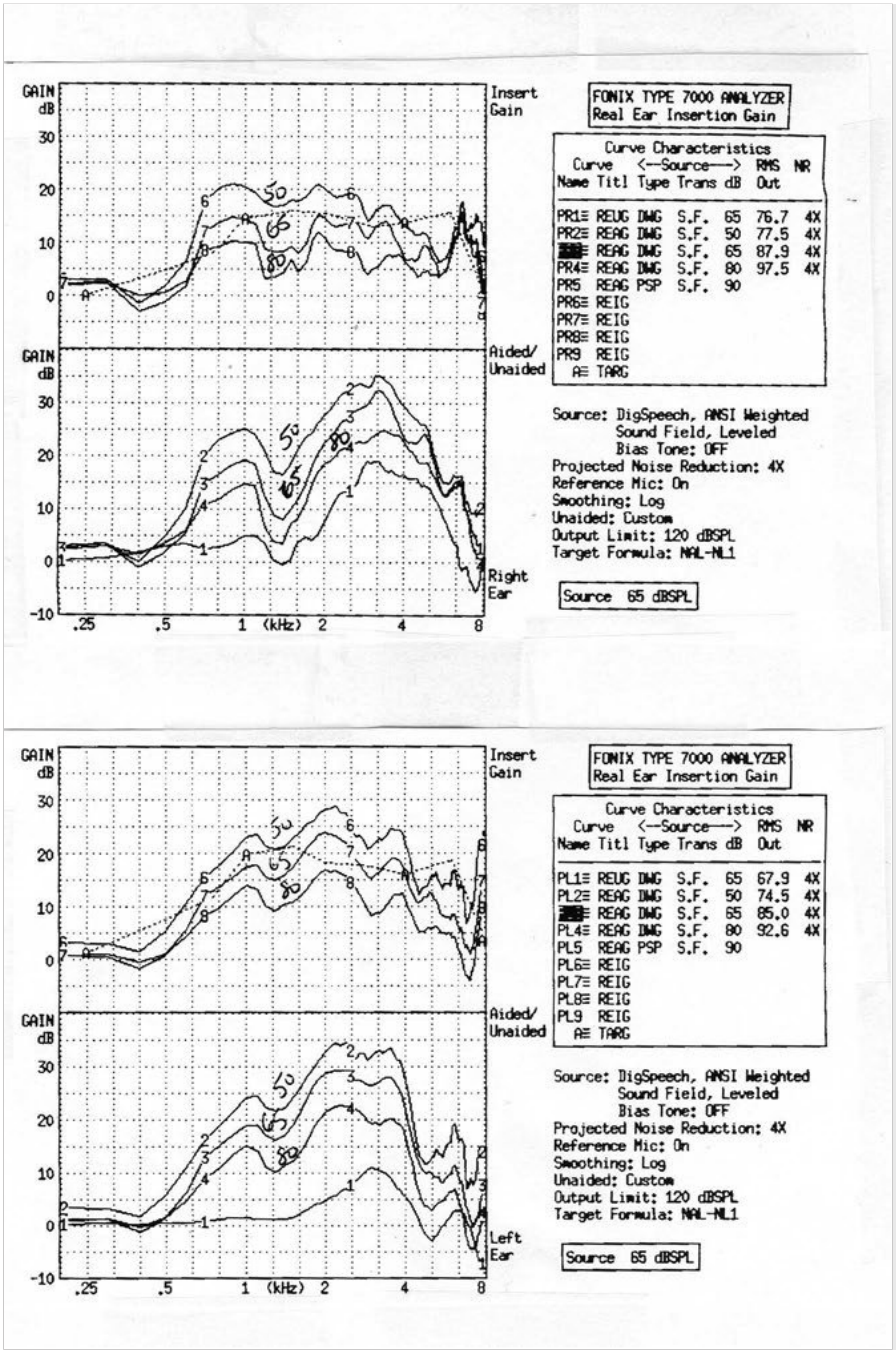
   GB was first counseled on the results of her audiometric examination indicating a flat mild to moderate sensorineural hearing loss. The WRSs were presented and discussed. It was explained that the words were presented at a suprathreshold level to allow for good audibility of speech sounds, and that, despite audibility of most speech sounds, approximately 50% of the words were repeated correctly in each ear. GB was counseled that even if a sound is presented clearly to her ears, she may not recognize the sound clearly. The poor WRSs were compared with a poorly designed loudspeaker. The sound may arrive at the loudspeaker clearly, but the poorly designed loudspeaker adds distortion. GB was also counseled regarding realistic expectations from amplification because sound clarity and speech recognition cannot be entirely restored through amplification.

   GB's WRSs were poorer than expected with a mild to moderate flat hearing loss. Pure-tone thresholds alone, however, cannot always accurately predict word recognition ability. Although audibility is important, signal and listener factors, including the listener's age, whether background noise is present, and the site of lesion, also affect speech recognition.
2. What would you recommend to the patient to improve communication?

   It was recommended that the patient continue use of her bilateral BTE hearing aids or consider new hearing aid technology. She was also counseled that her current hearing aids should be reprogrammed using real-ear probe microphone measurements to ensure that the hearing aids are providing appropriate audibility of soft sounds without overamplifying loud sounds. Finally, HATs, such as a personal FM system, were recommended to improve communication in noisy settings.
3. How can HAT improve the patient's communication compared with bilateral hearing aids alone?

   Hearing aids can incorporate directional microphone technology to help reduce background noise. Directional microphones are more sensitive to sound from the front than from the sides or back. Research has demonstrated that the use of directional microphones can improve speech recognition in noise by 2 to 8 dB, depending on the directional microphone design and the azimuth(s) of the loudspeakers emitting the noise.

   With a personal FM system, the transmitting FM microphone is usually close to the speaker's mouth. The speaker's speech is picked up by the microphone and because of its close proximity to the mouth, interference from reverberation, distance, and noise is minimal. The FM signal is then transmitted wirelessly from the microphone to the receiver worn by the listener. The receiver is available in two forms: a neckloop receiver worn around the listener's neck or audio boot receivers attached directly to BTE hearing aids. To use the neckloop receiver, the hearing aids must have a telecoil.



FONIX TYPE 7000 ANALYZER
Real Ear Insertion Gain

Curve Characteristics

Curve Name	Curve Titl	Source Type	Source Trans	Source dB	RMS Out	NR
PR1≡	REUG	DWG	S.F.	65	76.7	4X
PR2≡	REAG	DWG	S.F.	50	77.5	4X
[illegible]≡	REAG	DWG	S.F.	65	87.9	4X
PR4≡	REAG	DWG	S.F.	80	97.5	4X
PR5	REAG	PSP	S.F.	90		
PR6≡	REIG					
PR7≡	REIG					
PR8≡	REIG					
PR9	REIG					
A≡	TARG					

Source: DigSpeech, ANSI Weighted
Sound Field, Leveled
Bias Tone: OFF
Projected Noise Reduction: 4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source 65 dBSPL

FONIX TYPE 7000 ANALYZER
Real Ear Insertion Gain

Curve Characteristics

Curve Name	Curve Titl	Source Type	Source Trans	Source dB	RMS Out	NR
PL1≡	REUG	DWG	S.F.	65	67.9	4X
PL2≡	REAG	DWG	S.F.	50	74.5	4X
[illegible]≡	REAG	DWG	S.F.	65	85.0	4X
PL4≡	REAG	DWG	S.F.	80	92.6	4X
PL5	REAG	PSP	S.F.	90		
PL6≡	REIG					
PL7≡	REIG					
PL8≡	REIG					
PL9	REIG					
A≡	TARG					

Source: DigSpeech, ANSI Weighted
Sound Field, Leveled
Bias Tone: OFF
Projected Noise Reduction: 4X
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source 65 dBSPL

Fig. 66.2 Real-ear measures of the Exelia Art M hearing aids (Phonak).

The neckloop receiver sends the FM signal to the hearing aids via the telecoil. When hearing aids are coupled to an FM system, the hearing aids can usually be used in three settings: FM only (hearing aid microphones are off), FM plus hearing aid microphones, or hearing aid microphones alone. When in the FM only setting, very little background noise is audible to the listener. Lewis et al reported that FM systems in the FM only setting improved the signal to noise ratio (SNR) by 14 to 20 dB.

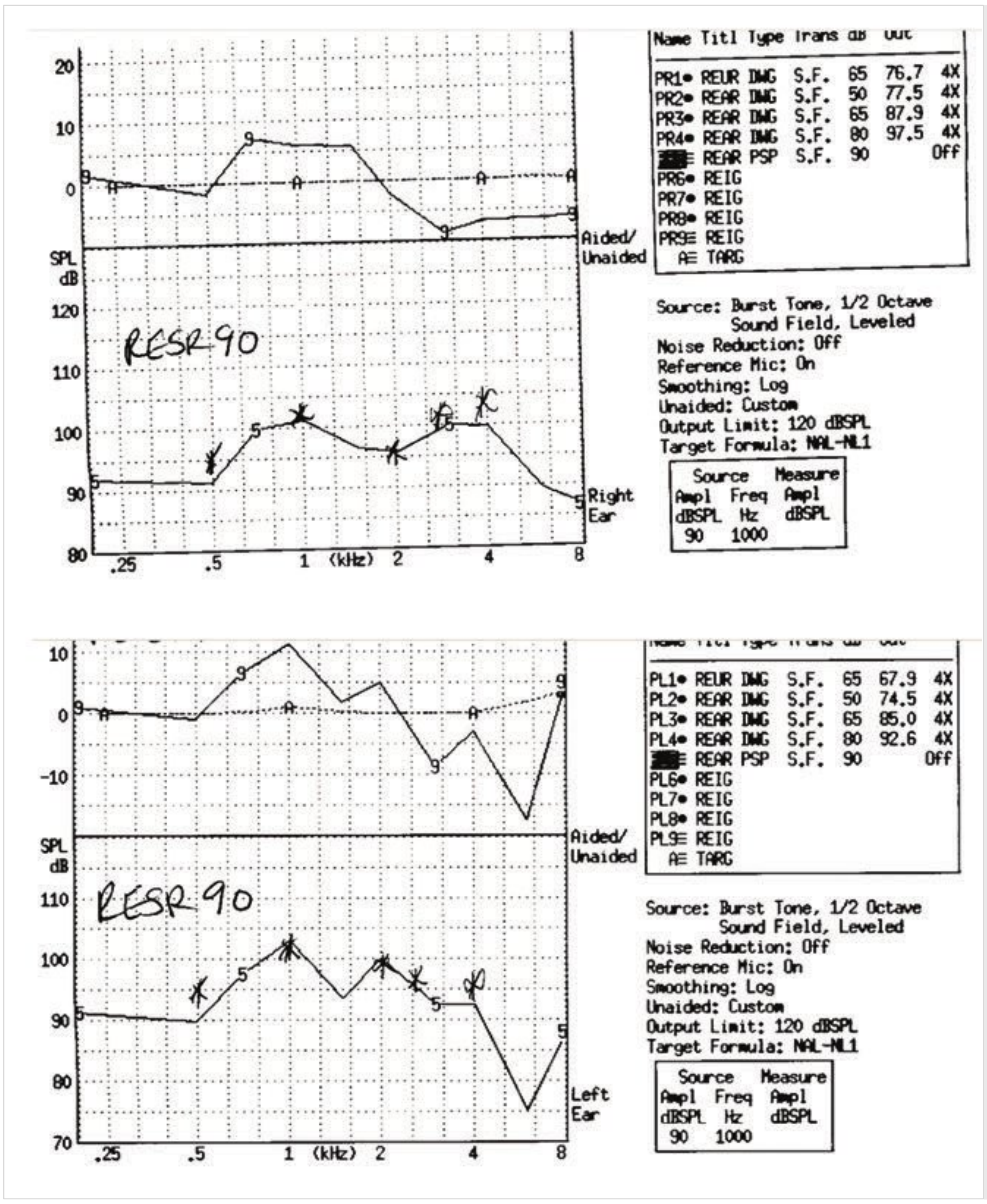
4. What type of verification is required to fit an FM system? The first step is to ensure that the hearing aids are working properly and programmed appropriately using real-ear measurements. All other measurements are based on the assumption that the hearing aid microphone response provides appropriate gain. Then, if a neckloop receiver is used, the telecoil response in the hearing aids should be measured to ensure proper function. This can be completed by measuring Sound Pressure Level in an inductive telephone simulator (SPLITS) in a hearing aid analyzer. In the Frye hearing aid analyzer (Frye Electronics, Inc., Tigard, OR), the Fonix telewand external magnetic simulator is used to measure the telecoil response. The SPLITS frequency response of the telecoil should be similar to the frequency response of the hearing aid microphone. A high-frequency average (HFA; average output at 1000, 1600, and 2500 Hz) is calculated from the SPLITS curve (HFA-SPLITS) and compared with the HFA of the hearing aid microphone's reference test gain (RTG) plus 60 dB SPL to calculate the relative simulated equivalent telephone sensitivity (RSETS). The RSETS value should be greater than or equal to 0 dB, indicating that the average gain at 1000, 1600, and 2500 Hz when the hearing aid is operating in the telecoil position is equal to or greater than when the

hearing aid is operating in the microphone position. An RSETS of greater than or equal to 0 dB ensures that the user will not perceive a decrease in loudness when switching from the hearing aid microphone to the telecoil.

After it has been demonstrated that the hearing aids and the telecoils (if using a neckloop receiver) are operating appropriately, FM transparency should be verified in the hearing aid analyzer. The output of the hearing aid in the microphone position is measured using a 65 dB SPL signal to measure the electroacoustic hearing aid response (EHA65). The FM setting is then activated in the hearing aid and the FM receiver is coupled to the hearing aid. If using a neckloop FM receiver, the FM setting in the hearing aid is the telecoil program. The transmitting microphone from the personal FM system is then placed in the hearing aid analyzer and the hearing aid, attached to the HA-2 2 cc coupler and test microphone, is placed on a foam pad on the top of the hearing aid analyzer. If using a neckloop receiver, the neckloop can be placed next to the hearing aid sitting on top of the hearing aid analyzer or the neckloop can be worn around the neck and the hearing aid can be held next to the ear during testing. The output of the hearing aid when communicating with the FM system is then measured. The same signal and signal level are used to measure the electroacoustic response of the hearing aid coupled to the FM system (EFMHA65). The EFMHA65 average at 750, 1000, and 2000 Hz should be no more than 2 dB less than the EHA65 average at 750, 1000, and 2000 Hz to confirm transparency. If the EFMHA65 average is more than 2 dB lower than the EHA65 average, then the FM settings should be adjusted for increased output.

66.5 Diagnosis and Recommended Treatment

After significant counseling and discussion of hearing aid technology and HAT, GB proceeded with new bilateral BTE hearing aids and an FM system. Bilateral Phonak Exelia Art M hearing aids with canal style Lucite earmolds were ordered. Real ear insertion gain (REIG) targets corrected for 20-channel summation and binaural summation were established using National Acoustic Laboratories' nonlinear fitting formula version 1 (NAL-NL1). The REIG was measured with input levels of 50, 65, and 80 dB SPL, and hearing aid programming was adjusted to ensure that the measured REIG was as close to prescribed REIG for NAL-NL1 as possible. Measurements revealed appropriate gain and smooth frequency response (Fig. 66.2, where the upper figure reports the right hearing aid and the lower figure



Name	Titl	Type	Trans	dB	Out	
PR1●	REUR	DWG	S.F.	65	76.7	4X
PR2●	REAR	DWG	S.F.	50	77.5	4X
PR3●	REAR	DWG	S.F.	65	87.9	4X
PR4●	REAR	DWG	S.F.	80	97.5	4X
▇≡	REAR	PSP	S.F.	90		Off
PR6●	REIG					
PR7●	REIG					
PR8●	REIG					
PR9≡	REIG					
A≡	TARG					

Source: Burst Tone, 1/2 Octave
Sound Field, Leveled
Noise Reduction: Off
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source		Measure
Ampl dBSPL	Freq Hz	Ampl dBSPL
90	1000	

PL1●	REUR	DWG	S.F.	65	67.9	4X
PL2●	REAR	DWG	S.F.	50	74.5	4X
PL3●	REAR	DWG	S.F.	65	85.0	4X
PL4●	REAR	DWG	S.F.	80	92.6	4X
▇≡	REAR	PSP	S.F.	90		Off
PL6●	REIG					
PL7●	REIG					
PL8●	REIG					
PL9≡	REIG					
A≡	TARG					

Source: Burst Tone, 1/2 Octave
Sound Field, Leveled
Noise Reduction: Off
Reference Mic: On
Smoothing: Log
Unaided: Custom
Output Limit: 120 dBSPL
Target Formula: NAL-NL1

Source		Measure
Ampl dBSPL	Freq Hz	Ampl dBSPL
90	1000	

Fig. 66.3 The real ear saturation response with an input level of 90 dB SPL (RESR90) measures.

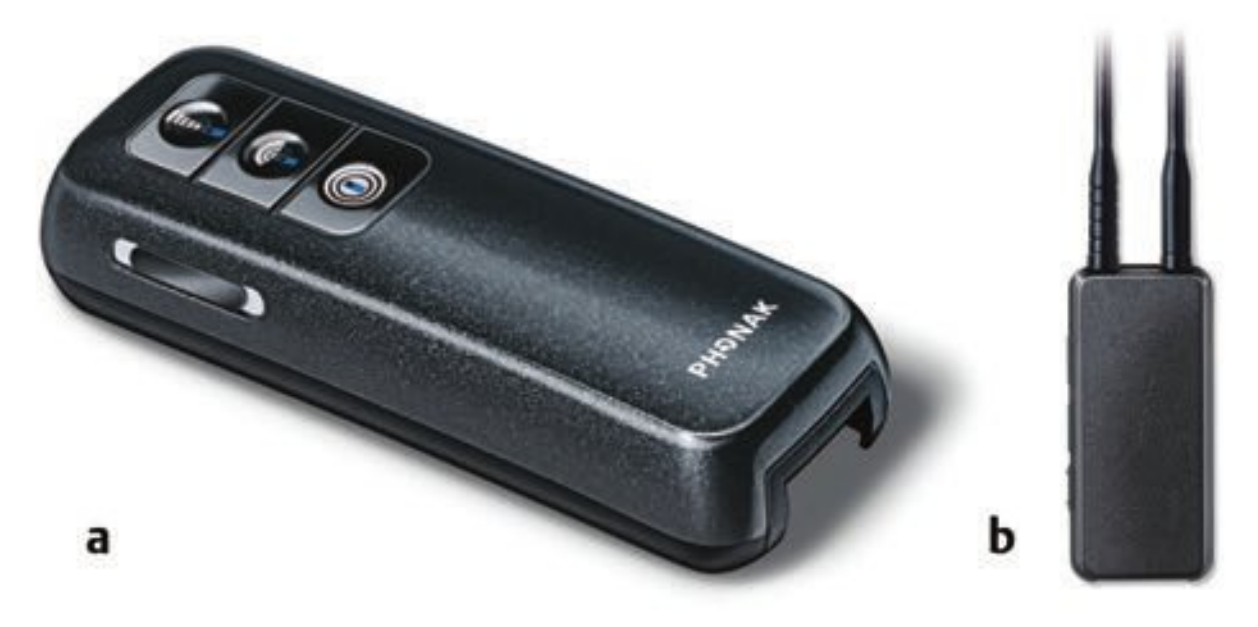
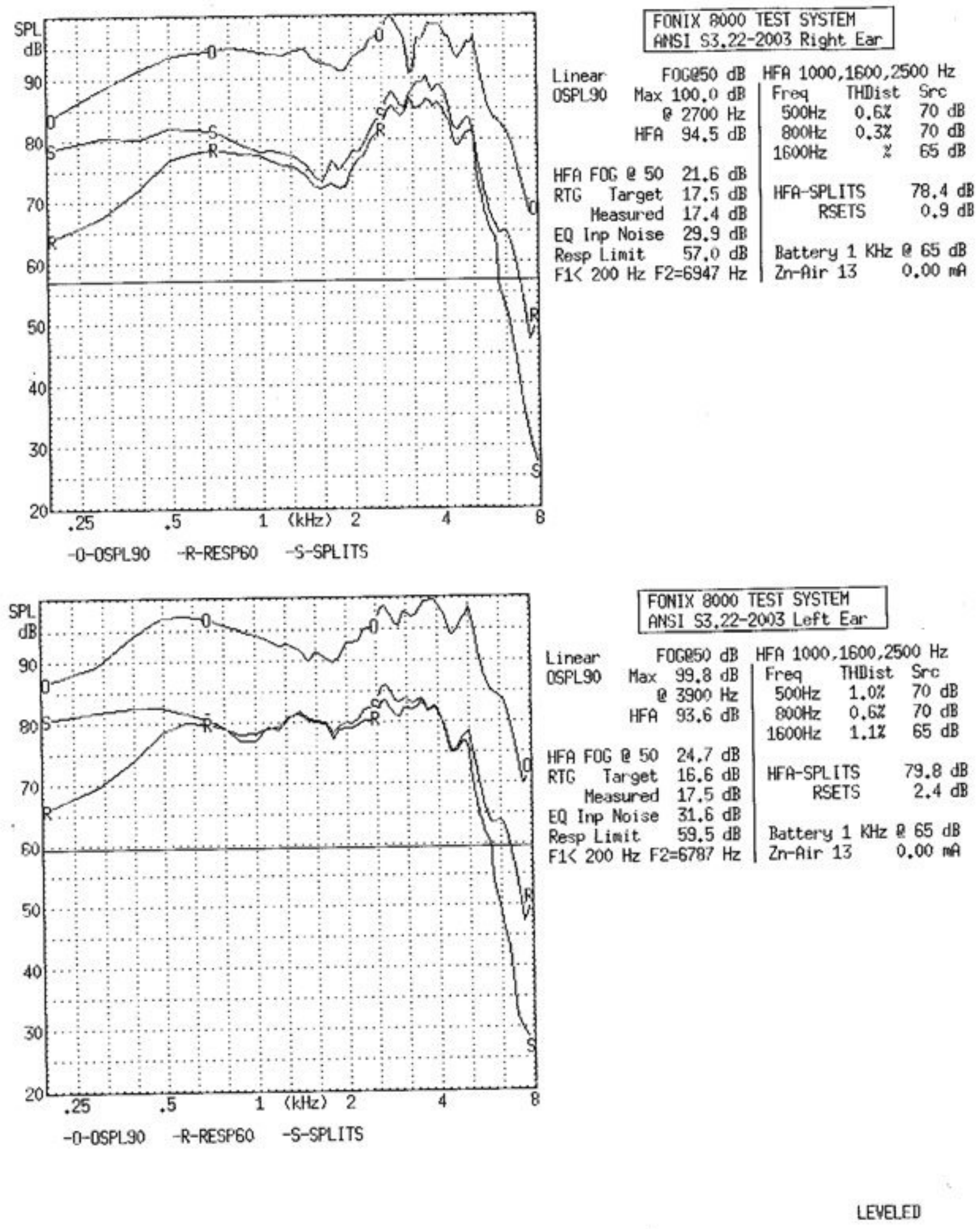


Fig. 66.4 The Phonak SmartLink + transmitter (a) and the Phonak MyLink + neckloop receiver (b). (Photos were retrieved from www.phonakpro.com and used with permission from Phonak.)

reports the left hearing aid). Curve 1 is real ear unaided gain (REUG), whereas curves 2 through 4 report the real ear aided gain (REAG) for input levels of 50 dB, 65 dB, and 80 dB SPL and curves 6 through 8 report the REIG for input levels of 50 dB, 65 dB, and 80 dB SPL. The "A" represents the NAL-NL1 target for an input level of 65 dB SPL. A modulated speech- weighted signal was used for all measurements.

The real ear saturation response (RESR90) was measured using a pure-tone sweep from 200 to 8000 Hz with an input level of 90 dB SPL (Fig. 66.3). The RESR90 curve corresponded to the appropriate frequency-specific dB SPL levels for loudness judgments of "loud, but OK," which were measured under headphones using the probe microphone prior to the hearing aid fit. In Fig. 66.3, the upper figure reports the right hearing aid, and the lower figure reports the left hearing aid. The dB SPL levels corresponding to the loudness judgments of "loud, but OK" for 500, 1000, 2000, 3000, and 4000 Hz are designated by the asterisks. Loudness judgments for composite speech noise presented at 50, 65, and 80 dB SPL were obtained. GB reported that an input of 50 dB SPL was "soft," 65 dB SPL was "comfortable," and 80 dB SPL was "loud, but OK."

Different FM systems were discussed and GB selected the Phonak SmartLink + transmitter (Fig. 66.4a), which can be paired to her Bluetooth cell phone, and the MyLink + neckloop receiver (Fig. 66.4b). Because a neckloop receiver was selected, the telecoils in the hearing aids were activated and the telecoil response was verified in the hearing aid analyzer (Fig. 66.5, where the upper figure reports measurements for the right hearing aid and the lower figure reports measurements on the left hearing aid). Curve "O" reports the output of the microphone using a pure-tone sweep at 90 dB SPL. Curve "R" reports the output of the microphone using a pure-tone sweep at 60 dB SPL, whereas curve "S" reports the output of the telecoil in response to a magnetic simulator The RSETS



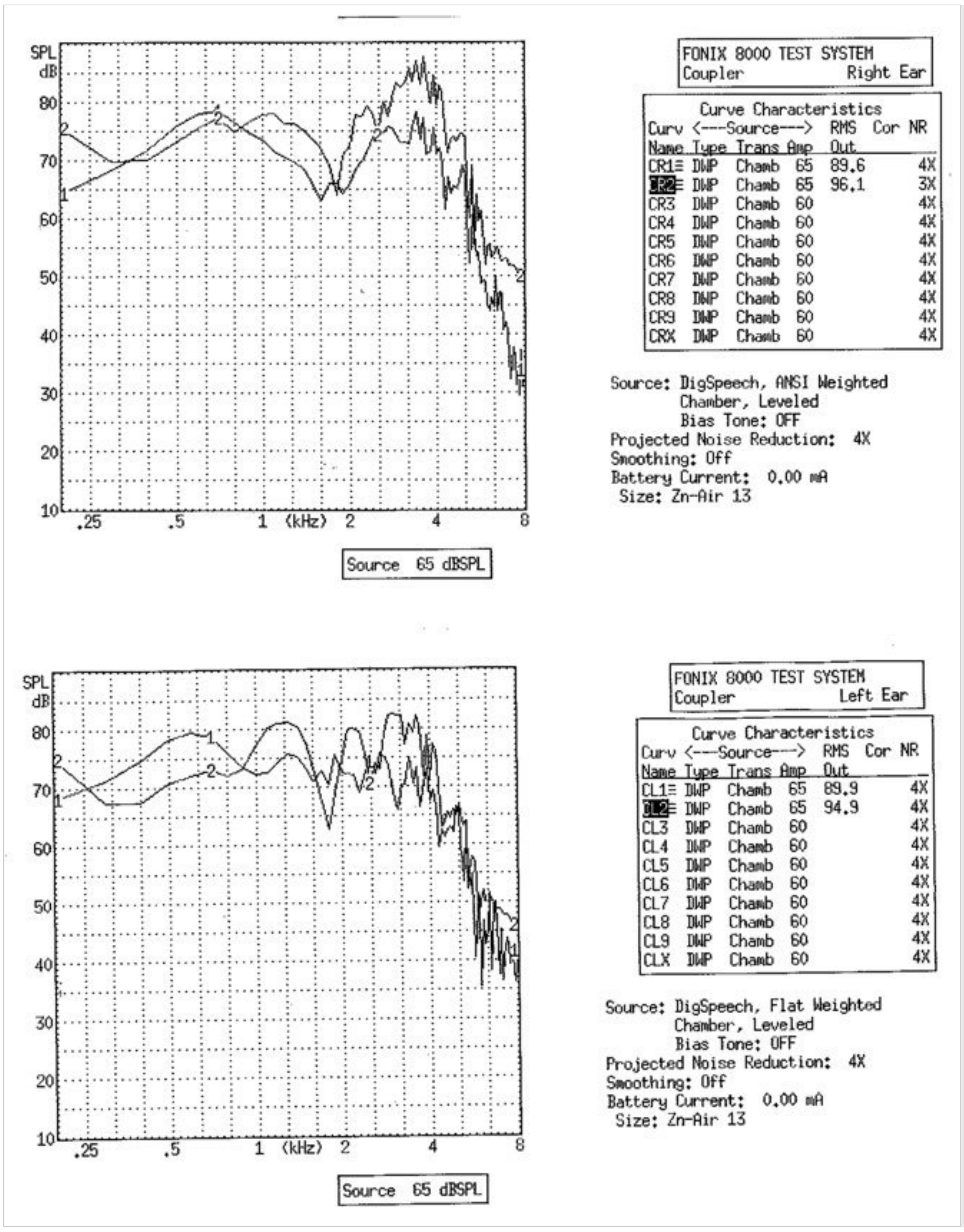
```
FONIX 8000 TEST SYSTEM
ANSI S3.22-2003 Right Ear

Linear        FOG@50 dB  HFA 1000,1600,2500 Hz
OSPL90    Max 100.0 dB | Freq     THDist  Src
             @ 2700 Hz |  500Hz    0.6%   70 dB
            HFA 94.5 dB|  800Hz    0.3%   70 dB
                       | 1600Hz      %    65 dB
HFA FOG @ 50   21.6 dB |
RTG    Target  17.5 dB | HFA-SPLITS      78.4 dB
     Measured  17.4 dB |      RSETS       0.9 dB
EQ Inp Noise   29.9 dB |
Resp Limit     57.0 dB | Battery 1 KHz @ 65 dB
F1< 200 Hz F2=6947 Hz  | Zn-Air 13       0.00 mA
```

```
FONIX 8000 TEST SYSTEM
ANSI S3.22-2003 Left Ear

Linear        FOG@50 dB  HFA 1000,1600,2500 Hz
OSPL90    Max  99.8 dB | Freq     THDist  Src
             @ 3900 Hz |  500Hz    1.0%   70 dB
            HFA 93.6 dB|  800Hz    0.6%   70 dB
                       | 1600Hz    1.1%   65 dB
HFA FOG @ 50   24.7 dB |
RTG    Target  16.6 dB | HFA-SPLITS      79.8 dB
     Measured  17.5 dB |      RSETS       2.4 dB
EQ Inp Noise   31.6 dB |
Resp Limit     59.5 dB | Battery 1 KHz @ 65 dB
F1< 200 Hz F2=6787 Hz  | Zn-Air 13       0.00 mA
```

Fig. 66.5 Verification of the telecoil.



```
FONIX 8000 TEST SYSTEM
Coupler              Right Ear

      Curve Characteristics
Curv <---Source---> RMS  Cor NR
Name Type Trans Amp Out
CR1≡ DWP  Chamb 65  89.6      4X
CR2≡ DWP  Chamb 65  96.1      3X
CR3  DWP  Chamb 60            4X
CR4  DWP  Chamb 60            4X
CR5  DWP  Chamb 60            4X
CR6  DWP  Chamb 60            4X
CR7  DWP  Chamb 60            4X
CR8  DWP  Chamb 60            4X
CR9  DWP  Chamb 60            4X
CRX  DWP  Chamb 60            4X

Source: DigSpeech, ANSI Weighted
        Chamber, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Smoothing: Off
Battery Current:  0.00 mA
 Size: Zn-Air 13
```

```
FONIX 8000 TEST SYSTEM
Coupler               Left Ear

      Curve Characteristics
Curv <---Source---> RMS  Cor NR
Name Type Trans Amp Out
CL1≡ DWP  Chamb 65  89.9      4X
CL2≡ DWP  Chamb 65  94.9      4X
CL3  DWP  Chamb 60            4X
CL4  DWP  Chamb 60            4X
CL5  DWP  Chamb 60            4X
CL6  DWP  Chamb 60            4X
CL7  DWP  Chamb 60            4X
CL8  DWP  Chamb 60            4X
CL9  DWP  Chamb 60            4X
CLX  DWP  Chamb 60            4X

Source: DigSpeech, Flat Weighted
        Chamber, Leveled
        Bias Tone: OFF
Projected Noise Reduction:  4X
Smoothing: Off
Battery Current:  0.00 mA
 Size: Zn-Air 13
```

Fig. 66.6 Verification of frequency-modulation (FM) transparency.

were 0.9 dB and 2.4 dB for the right and left hearing aids, respectively. The RSETS values were greater than 0 dB, and therefore GB should not perceive a decrease in loudness when switching from microphone to telecoil and neckloop FM receiver. Finally, the frequency response of the telecoil, curve "S," is very similar to the frequency response of the microphone, curve "R."

FM transparency was also measured (Fig. 66.6, where the upper figure reports the measures for the right hearing aid and the lower figure reports the left hearing aid). The EHA65 response is "1" and the EFMHA65 response is "2," and both curves reveal similar frequency responses. The averages at 750, 1000, and 2000 Hz were calculated for the EHA65 and EFMHA65 curves. As discussed earlier, the EFMHA65 average should be no more than 2 dB less than the EHA65 average to confirm transparency. For the right aid, the average for the EHA65 curve is approximately 72 dB SPL and the average for the EFMHA65 curve is approximately 76 dB SPL. The EFMHA 65 average is 4 dB greater than the EHA65 average, indicating FM transparency for the right aid. For the left aid, the average for the EHA65 curve is approximately 74 dB SPL and the average for the EFMHA65 curve is approximately 75 dB SPL. The EFMHA 65 average is 1 dB greater than the EHA65 average, indicating FM transparency for the left aid.

The hearing aids were programmed with hearing aid microphone only, telecoil only, and hearing aid microphone plus telecoil programs. GB was counseled on the use of the different programs, and the SmartLink+transmitter was also paired with her Bluetooth cell phone to allow for hands-free telephone use. Finally, GB was instructed on how to use the audio input cords of the FM system with her television at home.

66.6 Outcome

GB reported that her WRSs had never been explained in the past and that it helped explain many of the challenges she had been experiencing. She stated sounds can be loud, but still unclear. She also reported her new hearing aids were much clearer than her previous hearing aids, and she was hearing better with the new hearing aids alone than she had in the past. GB reported she was using the FM system in church, restaurants, when watching television, and when playing cards with her friends. She reported she was able to turn the captions off the television and she could watch some shows, such as the news, without using the captions or the FM system. The FM system, however, was very helpful for watching movies on her television. She also reported the new devices eased communication between her and her husband, and the new devices have allowed her to participate in activities that she had begun to shy away from, such as dining in restaurants and playing cards.

66.7 Key Points

1. Word recognition ability cannot always be predicted from the pure-tone thresholds, and, although audibility is important, other factors impact speech recognition.
2. Counseling on word recognition ability is very important and can be used to help explain many of the difficulties experienced by the patient.
3. FM technology can significantly improve speech recognition in noise compared with hearing aids alone.
4. Verification of the hearing aids and HATs is critically important. A clinician should not assume that the First-Fit rationale is appropriate or that HATs are functioning properly without verification.

Suggested Reading

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Part 7

Tinnitus Management

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67 Hearing Aids for Tinnitus

Judith Bird and David Baguley

A 72-year-old man with disabling tinnitus is referred to a tinnitus clinic.

67.1 Clinical History and Description

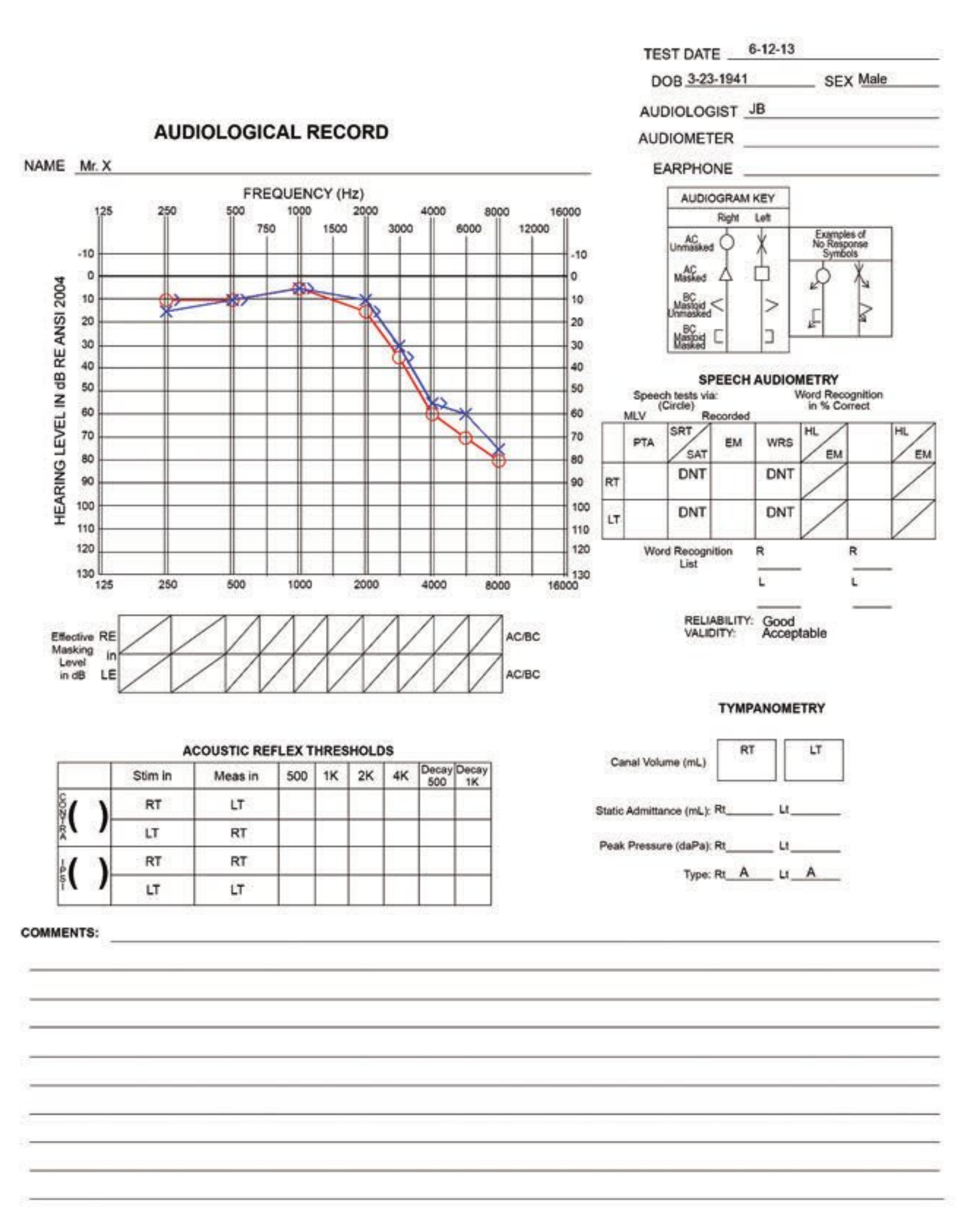
CE a 72-year-old man, was referred to a tinnitus clinic. CE reports troublesome tinnitus that has been present for more than 10 years. This is perceived as a "whistling in the center of his head." He has seen several ear, nose, and throat (ENT) specialists but derived minimal benefit. He is severely distressed by the tinnitus, his sleep is affected, and he reports that the tinnitus is "destroying his quality of life." CE was distressed and agitated during the consultation and admitted being depressed.

CE has a bilateral symmetrical mild to severe high-frequency sensorineural hearing loss at 3000 to 8000 Hz but reports no difficulty in hearing. He has previously tried hearing aids with no appreciable improvement to alleviate his symptoms. CE lives at home with his wife and describes hearing well at home during conversation, listening to the television, communicating on the telephone, hearing the doorbell. He is rarely engaged in social situations because he feels the tinnitus has negatively impacted his ability to enjoy life.

CE has a long history of exposure to artillery noise in the military for over 20 years without the use of hearing protection. He also reports a previous head injury where he lost consciousness in a traffic accident. His general physical health is otherwise good, although he appears depressed and anxious. He has been prescribed medication to help with sleeping (Zopiclone at a dose of 3.5 mg).

67.2 Audiological Testing

Audiological examination confirmed a mild to severe bilateral symmetrical high-frequency sensorineural hearing loss at 3000 to 8000 Hz with hearing within normal limits to 2000 Hz (Fig. 67.1). Tympanograms were type A bilaterally (normal



TEST DATE 6-12-13
DOB 3-23-1941 SEX Male
AUDIOLOGIST JB
AUDIOMETER
EARPHONE

AUDIOLOGICAL RECORD

NAME Mr. X

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV Recorded

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT		DNT		DNT			
LT		DNT		DNT			

Word Recognition List R L R L

RELIABILITY: Good
VALIDITY: Acceptable

TYMPANOMETRY

Canal Volume (mL) RT LT

Static Admittance (mL): Rt Lt
Peak Pressure (daPa): Rt Lt
Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim In	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA ( )	RT	LT						
	LT	RT						
IPSI ( )	RT	RT						
	LT	LT						

COMMENTS:

Fig. 67.1 Audiogram reporting bilateral high-frequency sensorineural hearing loss for CE.

middle ear pressure, compliance, and ear canal volume). Speech recognition testing was not completed because CE had no reports of any difficulty hearing. In addition to the audiological examination, the Tinnitus Handicap Inventory (THI) was completed in the presence of the clinician. CE's total score was 72% out of 100% indicating a severe handicap. Upon discussing the results, CE was relieved that finally someone believed him and was taking an interest. Despite the absence of CE's perceived hearing handicap as a result of his high-frequency hearing loss, the decision was taken to offer hearing aids as a treatment option. It was hoped that the hearing aids would provide some relief from the tinnitus.

67.3 Questions to the Reader

1. What is the evidence for using hearing aids as a treatment for tinnitus?
2. What extra care needs to be taken in programming hearing aids as a treatment for tinnitus?
3. How might the settings for a patient with tinnitus differ from those for a patient whose primary complaint was reduced hearing in background noise?
4. What other therapies/strategies are important in addition to hearing aids in patients with bothersome tinnitus?

67.4 Discussion of Questions to the Reader

1. What is the evidence for using hearing aids as a treatment for tinnitus?

   Hearing aids have been suggested as a treatment for tinnitus, and there are several mechanisms by which hearing aids may be successful. First, by providing increased awareness of environmental sounds, the hearing aids can reduce the "starkness" of the tinnitus by redirecting attention away from the tinnitus. In other words, due to increased awareness of genuine sounds, the intrusiveness of tinnitus may diminish. Furthermore, this increased sound enrichment provided by amplification may help partially mask the presence of the tinnitus. The increased stimulation to the auditory system may facilitate plastic reorganization of the central auditory system and thereby potentially facilitate habituation. By decreasing listening effort, hearing aids can also have a secondary effect of reducing stress that may be perpetuating a cycle of tinnitus generation. It is generally assumed that use of hearing aids should be combined with counseling for maximum tinnitus relief.

   Although they are widely used as a primary treatment for tinnitus for patients with a mild hearing loss, there is a paucity of robust evidence to support the use of hearing aids as a primary treatment for tinnitus alone. Also, for those few studies that have been published, few are well controlled. More recent studies report greater benefit with hearing aids, but it has been suggested that this is due to the improvements in technology with the introduction of digital hearing aids. Furthermore, Searchfield et al suggest that patients using hearing aids obtain greater benefit in relief from tinnitus than those patients receiving tinnitus counseling alone. Data from Marketrak VIII report greater benefit for tinnitus mitigation if best-practice protocols are followed. In this report, high best-practice ratings indicated that the service included aspects such as testing audiometric thresholds and speech audiometry in a sound-treated booth and providing validation and verification of the hearing aid fitting. This suggests that not only the technology but also the method and quality of the fitting of the hearing aids are important factors. McNeill et al indicate that the benefit of hearing aids for relief from tinnitus is greater when the pitch of the tinnitus falls within the frequency range of the hearing aids. This conclusion suggests that the benefits of extended-range high-frequency hearing aids may be of interest, though the evidence for this is equivocal at present.
2. What extra care needs to be taken in programming hearing aids as a treatment for tinnitus?

   Some patients with tinnitus have increased sensitivity to loud sounds, so care must be taken when programming the hearing aids to ensure that loud sounds do not exceed tolerance levels. While testing for uncomfortable loudness levels (UCLs) may be contraindicated due to a patient's sensitivity to loud sounds, careful setting of the maximum output of hearing aids for louder sounds should be completed in the clinic. At the very least, tolerance to sounds can be examined subjectively using familiar environmental sounds such as a raised voice or rattling utensils. Tolerance can be assessed more accurately during real ear measurement (REM) testing with a standardized signal (e.g., International Speech Test Signal at 80 dB sound pressure level [SPL]). The patient needs to leave the clinic confident that the hearing aid can be used comfortably in real-life listening situations. In extreme cases, optimal amplification may need to be sacrificed so that usable amplification of soft sounds can be achieved.
3. How might the settings for a patient with tinnitus differ from those for a patient whose primary complaint was reduced hearing in background noise?

   Adaptive directional microphones and noise reduction algorithms are used to optimize hearing and maximize listening comfort in background noise. These features, however, also lead to the reduction of environmental sound that may help reduce tinnitus awareness. Because of this, hearing aids with an omnidirectional microphone and no noise reduction are preferable. A low compression kneepoint (e.g., < 50 dB SPL input level) will help to maximize audibility of soft environmental noises. Use of soft-squelch or expansion circuits should be avoided because these features are designed to reduce the amplification for very soft sounds that are usually not desirable. Although these differences imply that use of a "tinnitus program" may be beneficial, care should be taken to avoid any counseling that effectively cues the patient to listen for the tinnitus (such as counseling a patient to change the hearing program when the tinnitus is worse) because this may serve to perpetuate symptoms. Adjusting programs for "quiet situations" (hearing aid settings optimized for tinnitus reduction) and "noisy situations" (settings optimized for speech clarity) may provide more helpful terminology. Similarly, although user-controlled volume control (VC) can provide a greater sense of control, this type of VC can serve to maintain the focus on the patient's hearing and hence the tinnitus. In many cases, an automatic VC may be preferable.

4. What other therapies/strategies are important in addition to hearing aids in patients with bothersome tinnitus?
For patients with bothersome tinnitus and hearing loss, it is important that they receive counseling in addition to other treatment strategies such as amplification. A recent randomized controlled trial indicated that embedding a hearing aid fitting (or sound generators) in combination with patient education and counseling better managed the fear that a patient may experience, which led to significantly better long-term outcome.

67.5 Diagnosis and Recommended Treatment

CE was seen by a consultant audiologist in the tinnitus clinic where he was able to discuss the impact of his disabling tinnitus. It was very important that CE's general physician was fully involved in the progress of his treatment to monitor his general and psychological well-being given the extent and seriousness of CE's depression. Counseling was provided using appropriate terms to help CE understand the mechanisms that cause tinnitus. CE was given sound enrichment devices to help reduce his awareness of the tinnitus. Specifically CE found some relief from a bedside digital environmental sound generator that enabled him to achieve sleep more easily.

At his follow-up visit, CE reported experiencing some relief but was still experiencing disabling tinnitus. He was encouraged to consider a hearing aid trial despite his minimal complaint of hearing difficulty. CE was fitted with bilateral digital behind-the-ear hearing aids. Slim-tubes and open domes were used to optimize natural low-frequency hearing and minimize occlusion of the ear. Given the minimal impact of the hearing loss, hearing aids were programmed to provide maximum tinnitus relief. To accomplish this, compression was programmed to maximize audibility of soft sounds with compression kneepoints at approximately 40 dB SPL. Noise reduction was turned off, and the microphone was programmed for omnidirectional performance. Because these adjustments may not be optimal for maximizing speech recognition, CE was provided a second listening program with settings optimized for speech recognition (e.g., fit to National Acoustic Laboratories' nonlinear fitting formula version 1 [NAL-NL1], noise reduction on, and adaptive directionality turned on), which he was instructed to use in situations where it was important for him to recognize speech as clearly as possible.

67.6 Outcome

CE returned for a follow-up visit reporting that the hearing aids were very helpful. He stated that the hearing aids "absorb the tinnitus" and provide 95%inhibition. Datalogging confirmed his report that he was using his hearing aids for the entire day. The hearing aids had been used almost exclusively on the first listening program to maximize environmental awareness. CE managed his sleep by using sound enrichment from either a radio or a bedside noise generator (using sound of the sea) and sleeping tablets (Zopiclone) prescribed by his general practitioner. His THI score had decreased to 24% indicating that, although the tinnitus was occasionally troublesome, his quality of life had significantly improved.

67.7 Key Points

1. In combination with counseling and sound enrichment, hearing aids can provide significant benefit for individuals with disabling tinnitus. This appears to be true even when the hearing loss is mild and self-reported hearing difficulty is negligible
2. Effectiveness of hearing aids in reducing tinnitus awareness can be maximized by careful programming of the hearing aid (s), notably by optimizing gain for soft sounds (i.e., using a low compression kneepoint) and turning off noise reduction and directional microphones.
3. Care should be taken in providing a separate tinnitus program because it may serve to act as a cue to the patient to listen for the tinnitus.

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68 Headaches, Hyperacusis, and Tinnitus

Diane Duddy

A 31-year-old female presents with headaches, hyperacusis, and tinnitus triggered by electrical emissions.

68.1 Clinical History and Description

A 31-year-old female was referred to the audiology clinic by an otologist due to the patient report of headaches, tinnitus, and hyperacusis related to a hypersensitive central nervous system. The patient reported constant bilateral high-pitched "ringing" and "buzzing" tinnitus for approximately 6 years. Eighteen months prior to the referral, the patient began experiencing severe daily headaches, which were controlled by medication (Lyrica, topiramate, and hydrocodone) prescribed by the patient's neurologist. Three months prior to the otology referral the patient noticed that sounds such as projector fans, public address systems, automobile air conditioning fans, and fluorescent lights triggered the uncontrollable headaches and increased the tinnitus. She also reported that these episodes of headaches and tinnitus interfered with her concentration. She denied any circumstances or changes in her health history associated with the increased headaches and tinnitus.

The patient had a history of occasional recreational noise exposure through attending loud concerts and recreational shooting. These experiences, however, did not reportedly correlate with her otologic/neurologic symptoms. She also reported a significant history of occupational noise exposure in a manufacturing environment. Occasionally, she was working on the plant floor and reported using earplug hearing protection as mandated by her employer. She reported the earplugs reduced the headaches, sensitivity to sound, and tinnitus in a noisy environment. In a quiet office, however, the headaches were severe. Just prior to the consult, the patient moved her office closer to a heating, ventilation, and air conditioning (HVAC) unit located outside the building. When the HVAC was operating she reported diminished tinnitus and hypersensitivity and fewer headaches. She believed her tinnitus, hyperacusis, and headaches were correlated with the electrical emissions from various sources she sensed, felt, or heard in a quiet environment. In a quiet office or home, turning on fluorescent lights, computers, DVD players, television, or high-efficiency light bulbs increased her headaches, hypersensitivity, and tinnitus. Due to the quiet environment required for sleeping, the increased tinnitus prevented her from falling asleep and awakened her, on average, four times per night. The tinnitus and headaches were affecting her work, social, and home life. She had difficulty performing her job, and she could no longer complete her housework or play with her children. She began to withdraw socially because she was uncertain as to what would evoke the headaches. She was also being treated unsuccessfully with steroid injections at a pain clinic in an attempt to relieve the severity of her headaches. As a result of her symptoms, two magnetic resonance imaging (MRI) scans, one of her cervical spine (with and without contrast) and one of her brain and brainstem (with and without contrast) as well as a magnetic resonance angiography (MRA) scan were performed and were normal.

68.2 Audiological Testing

A comprehensive audiological evaluation revealed that pure-tone thresholds, speech recognition thresholds (SRTs), word recognition scores (WRSs), and immittance audiometry were all normal.

68.3 Questions to the Reader

1. Is the referral to audiology for headaches/tinnitus/hyperacusis management appropriate?
2. What additional audiological measurements would be recommended?
3. What treatment options might be considered?

68.4 Discussion of Questions to the Reader

1. Is the referral to audiology for headaches/tinnitus/hyperacusis management appropriate?
   Yes, the referral was appropriate because the primary source of her symptoms cannot be determined, and treatments for her headaches had not been successful in alleviating her tinnitus, hyperacusis, or headaches. It could not be determined if the headaches exacerbated the tinnitus and hyperacusis or the tinnitus and hyperacusis exacerbated the headaches.
   Hyperacusis is commonly defined, among audiology professionals specializing in treatment of patients with tinnitus and hyperacusis, as a subjective decreased tolerance for sounds that would otherwise be tolerable to individuals with normal hearing. Reducing the patient's hyperacusis or hypersensitivity to sound as well as tinnitus awareness and disturbance may reduce the severity and frequency of her headaches. As was reported earlier in the clinical history and description, the patient reported her hypersensitivity was reduced by the presence of other sound. If the treatment by audiology was successful, a program administered by her neurologist to monitor and reduce her medication for her headaches may result in an improvement in the patient's ability to concentrate (see adverse drug reactions).
2. What additional audiological measurements are recommended?
   Additional audiological measures could include the following:
   a) Loudness discomfort levels (LDLs in dB HL) to measure and monitor hyperacusis outcomes
   b) Subjective ratings to evaluate her perceived awareness and disturbance from her tinnitus
   c) Standardized subjective tinnitus measurements such as the Tinnitus Handicap Inventory, Tinnitus Functional

Index, or Tinnitus Reaction Questionnaire to measure and monitor the patient's disturbance or handicap from the tinnitus

d) Subjective ratings, to evaluate her perceived tolerance to electrical emission sounds associated with triggering headaches, hyperacusis and tinnitus, no known scale or questionnaire are presently available.

e) A tinnitus assessment consisting of tinnitus pitch and loudness matching may be considered. At this time, however, measurement of the patient's tinnitus pitch, loudness, maskability, and residual inhibition provides no guideline for treatment or outcome measures.

3. What treatment options might be considered?
   Based on Jastreboff's recommendations, hyperacusis should be addressed prior to tinnitus management and treated with a systematic program of sound therapy. The treatment protocol recommends a gradual increasing exposure to sounds such as pink noise or white noise using a personal listening device (PLD) or ear-level sound generators. The gradual increase in sound exposure depends on the patient's sensitivity to sound. This process may begin with the patient wearing the devices with the volume control (VC) in the off position for several days, followed by wearing the devices with the VC turned on and adjusted to an inaudible level for several more days before gradually increasing the VC to a comfortable listening level (CLL). Also, based on the patient's sensitivity to sound, the amount of time the devices are worn may need to be gradually introduced. Sound therapy protocol suggests the patient would wear the devices at a CLL for 8 or more hours per day. Options should be demonstrated and provided on a trial basis in order for the patient to determine if the recommended sounds affect her headaches.

68.5 Additional Testing

Initial LDLs (dB HL) were measured using the Neuromonics, Inc. (Westminster, CO) protocol for measuring LDLs. LDLs were measured separately for each ear (Table 68.1). The right ear LDLs for 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz indicate the LDLs are below 100 dB HL; the left ear LDLs for 500 Hz, 1000 Hz, 2000 Hz, and 4000 Hz also indicate that the LDLs are below 95 dB HL. According to recent studies, LDLs for adults with normal hearing are 95 to 100 dB HL. Therefore, LDLs below 95 dB HL would suggest sound tolerance problems.

Baseline subjective tinnitus ratings, based on the Neuromonics, Inc., protocol were obtained from the patient. This protocol asks two questions, "What percentage of your awake time in the past week have you been aware of your tinnitus?" and "What percentage of your awake time in the past week have you been disturbed or bothered by your tinnitus?" These baseline responses are used to compare the patient's responses at subsequent consultations to evaluate progress. A significant improvement is considered a 40% or greater reduction in the patient's tinnitus awareness as well as her tinnitus disturbance or bother. This patient's responses at the initial evaluation were 100% tinnitus awareness during her awake time in the past week and 80%tinnitus disturbance or bother during her awake time in the past week.

Table 68.1 Loudness discomfort levels (LDLs) (dB HL) measured at the initial tinnitus and hyperacusis consult

LDLs at (dB HL) at Initial Visit				
	500 Hz	1000 Hz	2000 Hz	4000 Hz
Right	72	70	74	76
Left	72	76	70	72

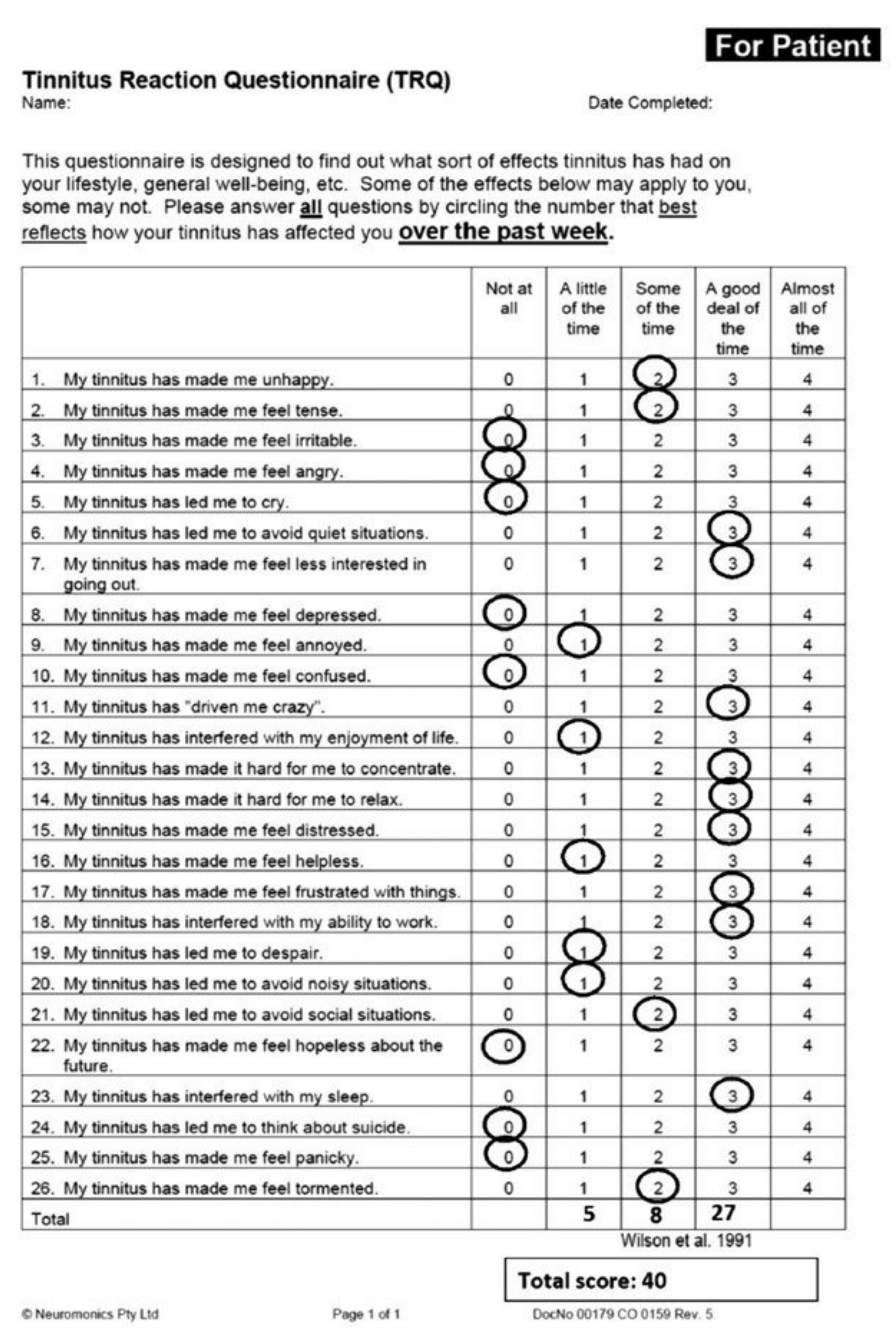
The Tinnitus Reaction Questionnaire (TRQ) was administered and the patient's total score was 40 (Fig. 68.1). The total possible score on the TRQ is 104 points and a score above 17 indicates clinically significant distress from tinnitus. Question 24 pertains to thoughts of suicide and a score higher than 0 indicates a referral should be made to a mental health professional for further evaluation. This patient's response on question 24 was 0.

68.6 Additional Questions to the Reader

1. What factors should be considered when measuring LDLs in hyperacusis patients?
2. What referrals and additional testing may be appropriate for this patient?

68.7 Discussion of Additional Questions to the Reader

1. What factors should be considered when measuring LDLs in hyperacusis patients?
   For treating this patient, intrasubject reliability for test–retest of LDL measures would be important as an evaluation of outcome of treatment. Studies have reported that intrasubject test–retest reliability differences are usually less than 5 dB for LDL measures. The Neuromonics, Inc., protocol used by this clinic provides written instructional wording to assure consistency in patient task instructions as well as specific guidelines for the audiologist's LDL measure procedure.
2. What additional referrals may be appropriate for this patient?
   Patients reporting tinnitus and hyperacusis may benefit by referral to a mental health professional. If patient comments or behaviors, initial intake tinnitus and hyperacusis history form, or tinnitus questionnaires suggest anxiety or depression, a referral to a mental health professional may facilitate audiological treatment outcomes. There are several mental health screening tools such as the Patient Health Questionnaire depression module (PHQ-9) available to audiologists. Any response suggesting thoughts of suicide warrants immediate referral to a mental health professional or physician. Additionally, if the patient's initial tinnitus and hyperacusis history form or tinnitus questionnaire suggest sleep-related problems such as difficulty falling asleep, frequent awakening, or insufficient sleep associated with tinnitus or hyperacusis, a referral to a mental health professional may be beneficial.



For Patient

Tinnitus Reaction Questionnaire (TRQ)

Name: Date Completed:

This questionnaire is designed to find out what sort of effects tinnitus has had on your lifestyle, general well-being, etc. Some of the effects below may apply to you, some may not. Please answer **all** questions by circling the number that best reflects how your tinnitus has affected you **over the past week.**

	Not at all	A little of the time	Some of the time	A good deal of the time	Almost all of the time
1. My tinnitus has made me unhappy.	0	1	[✓ 2]	3	4
2. My tinnitus has made me feel tense.	0	1	[✓ 2]	3	4
3. My tinnitus has made me feel irritable.	[✓ 0]	1	2	3	4
4. My tinnitus has made me feel angry.	[✓ 0]	1	2	3	4
5. My tinnitus has led me to cry.	[✓ 0]	1	2	3	4
6. My tinnitus has led me to avoid quiet situations.	0	1	2	[✓ 3]	4
7. My tinnitus has made me feel less interested in going out.	0	1	2	[✓ 3]	4
8. My tinnitus has made me feel depressed.	[✓ 0]	1	2	3	4
9. My tinnitus has made me feel annoyed.	0	[✓ 1]	2	3	4
10. My tinnitus has made me feel confused.	[✓ 0]	1	2	3	4
11. My tinnitus has "driven me crazy".	0	1	2	[✓ 3]	4
12. My tinnitus has interfered with my enjoyment of life.	0	[✓ 1]	2	3	4
13. My tinnitus has made it hard for me to concentrate.	0	1	2	[✓ 3]	4
14. My tinnitus has made it hard for me to relax.	0	1	2	[✓ 3]	4
15. My tinnitus has made me feel distressed.	0	1	2	[✓ 3]	4
16. My tinnitus has made me feel helpless.	0	[✓ 1]	2	3	4
17. My tinnitus has made me feel frustrated with things.	0	1	2	[✓ 3]	4
18. My tinnitus has interfered with my ability to work.	0	1	2	[✓ 3]	4
19. My tinnitus has led me to despair.	0	[✓ 1]	2	3	4
20. My tinnitus has led me to avoid noisy situations.	0	[✓ 1]	2	3	4
21. My tinnitus has led me to avoid social situations.	0	1	[✓ 2]	3	4
22. My tinnitus has made me feel hopeless about the future.	[✓ 0]	1	2	3	4
23. My tinnitus has interfered with my sleep.	0	1	2	[✓ 3]	4
24. My tinnitus has led me to think about suicide.	[✓ 0]	1	2	3	4
25. My tinnitus has made me feel panicky.	[✓ 0]	1	2	3	4
26. My tinnitus has made me feel tormented.	0	1	[✓ 2]	3	4
Total		5	8	27	

Wilson et al. 1991

Total score: 40

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Fig. 68.1 Tinnitus Reaction Questionnaire completed at the initial tinnitus and hyperacusis consultation.

68.8 Diagnosis and Recommended Treatment

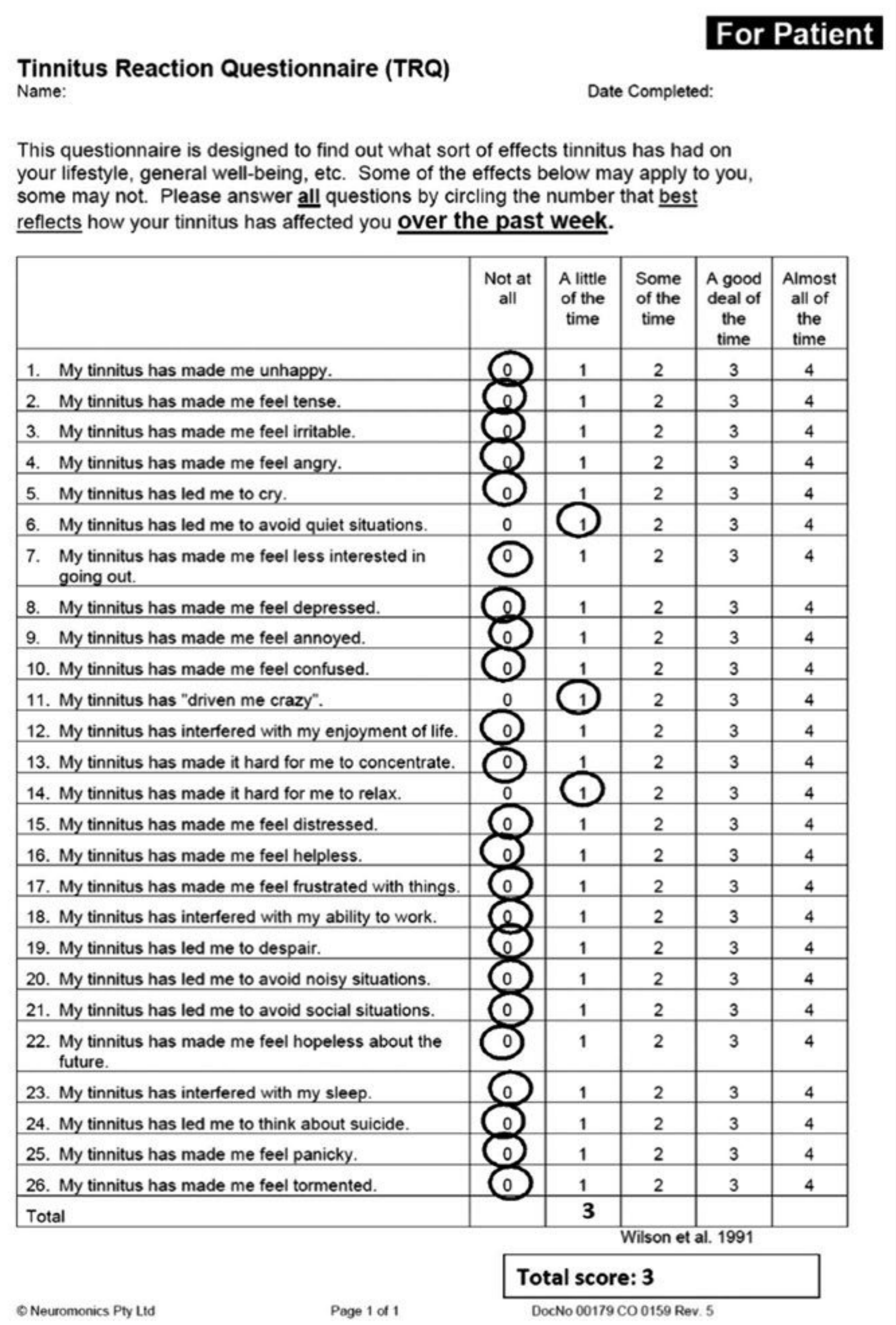
The patient was diagnosed with hyperacusis and subjective tinnitus. A PowerPoint presentation prepared by the author for the purpose of counseling tinnitus and hyperacusis patients was presented and discussed with the patient and her family. This presentation includes information on the neurophysiological principles of tinnitus and hyperacusis, current theories of increased central gain, caution on overuse of hearing protection, current tinnitus and hyperacusis research, as well as treatment goals and options. Sound therapy using white or pink noise was recommended with the option of using a PLD such as an MP3 or iPod, tabletop device, or sound generator. Sound therapy using either white or pink noise, whichever the patient prefers, involves the gradual introduction of comfortable output levels of the selected noise, which are then used for at least 8 hours every day. Improvements of LDLs are generally expected in 6 months. White noise and pink noise used with a PLD or tabletop device are available on the Internet. A demonstration of sound generators was provided. Sound generators are currently available as custom in-the-ear (ITE) or thin-tube behind-the-ear (BTE) devices. The custom devices are made from an ear impression and designed to allow for as little occlusion as possible, thus allowing the patient to hear communication with no interference from the physical device. The patient initially used a PLD with white noise uploaded from the Internet because most of her workday was in an office environment. She found the PLD helpful; however, it was not practical to use at home, work, or social situations that required communication. Subsequently, the patient was fitted bilaterally with General Hearing Instruments, Inc. (New Orleans, LA) custom ITE white-noise sound generators. The recommended protocol for using a sound generator for hyperacusis treatment was to begin with low-level sound and gradually increase the output to a CLL. It was recommended that the patient wear the devices for at least 8 hours a day. In addition, the patient and her husband were provided with recommended readings, including the Consumer Handbook on Tinnitus (Richard S. Tyler) to facilitate the patient's understanding of hyperacusis and tinnitus. A 30-day trial period was provided for the patient to determine if the use of the sound generators exacerbated her headaches or tinnitus. The patient reported that with guidance from her neurologist she would begin reducing her medications for her headaches once she acclimated to the sound generators.

68.9 Outcome

At a follow-up office visit 2 weeks following the fitting, the patient reported the sound generators seemed to be helping and did not exacerbate her headaches. Eleven months after treatment began, the patient reported wearing the sound generators consistently throughout the day at a CLL for 7 months, and given the improvements in her headaches and tinnitus, was now wearing the sound generators 30 or 60 minutes per day depending on her environment. The patient's LDLs were remeasured and found to be greater than 95 dB HL at 500 Hz, 1000 Hz, and 4000 Hz bilaterally (Table 68.2) indicating significant improvement in loudness tolerance. Her TRQ score was reduced to 3 (Fig. 68.2). Scores below 17 on the TRQ indicate

Table 68.2 Loudness discomfort levels (LDLs) (dB HL) measured at the conclusion of sound therapy

LDLs (dB HL) at Final Visit				
	500 Hz	1000 Hz	2000 Hz	4000 Hz
Right	96	96	94	98
Left	94	96	92	96



For Patient

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Date Completed:

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	Not at all	A little of the time	Some of the time	A good deal of the time	Almost all of the time
1. My tinnitus has made me unhappy.	[✓ 0]	1	2	3	4
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3. My tinnitus has made me feel irritable.	[✓ 0]	1	2	3	4
4. My tinnitus has made me feel angry.	[✓ 0]	1	2	3	4
5. My tinnitus has led me to cry.	[✓ 0]	1	2	3	4
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12. My tinnitus has interfered with my enjoyment of life.	[✓ 0]	1	2	3	4
13. My tinnitus has made it hard for me to concentrate.	[✓ 0]	1	2	3	4
14. My tinnitus has made it hard for me to relax.	0	[✓ 1]	2	3	4
15. My tinnitus has made me feel distressed.	[✓ 0]	1	2	3	4
16. My tinnitus has made me feel helpless.	[✓ 0]	1	2	3	4
17. My tinnitus has made me feel frustrated with things.	[✓ 0]	1	2	3	4
18. My tinnitus has interfered with my ability to work.	[✓ 0]	1	2	3	4
19. My tinnitus has led me to despair.	[✓ 0]	1	2	3	4
20. My tinnitus has led me to avoid noisy situations.	[✓ 0]	1	2	3	4
21. My tinnitus has led me to avoid social situations.	[✓ 0]	1	2	3	4
22. My tinnitus has made me feel hopeless about the future.	[✓ 0]	1	2	3	4
23. My tinnitus has interfered with my sleep.	[✓ 0]	1	2	3	4
24. My tinnitus has led me to think about suicide.	[✓ 0]	1	2	3	4
25. My tinnitus has made me feel panicky.	[✓ 0]	1	2	3	4
26. My tinnitus has made me feel tormented.	[✓ 0]	1	2	3	4
Total		3			

Wilson et al. 1991

Total score: 3

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Fig. 68.2 Tinnitus Reaction Questionnaire completed at the completion of the tinnitus and hyperacusis sound therapy.

no significant tinnitus disturbance. Her subjective response to her tinnitus awareness in the past week was reduced to 10% of her awake time, and her tinnitus disturbance in the past week was reduced to 5% of her awake time. A comprehensive audiological examination revealed all results were within normal limits. The patient reported occasionally forgetting to wear the sound generators, but she wears the sound generators when she knows there is a public address system in the room or electrical equipment emitting high-pitched sounds. The tinnitus no longer interferes with her sleep and does not awaken her at night. She no longer has headaches from sound sources as long as she wears the sound generators in the situations described above. In addition, she is no longer taking headache medications.

68.10 Key Points

1. Hypersensitivity to acoustic stimuli may be treated successfully with sound therapy and counseling.
2. When possible testing and treatment outcomes measurements should be consistent and standardized.

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69 Somatic Tinnitus

Craig W. Newman and Sharon A. Sandridge

A 57-year-old female was referred for left-sided tinnitus and otalgia (ear pain).

69.1 Clinical History and Description

BN, a 57-year-old female, was referred to the tinnitus management clinic (TMC) at an audiology clinic. The TMC provides a comprehensive approach to the evaluation and management of patients with tinnitus through a multidisciplinary team (audiology, dentistry, neurology, physical therapy, and psychology) focused on counseling/education and the provision of individualized management recommendations following screenings (not comprehensive evaluations) conducted by each specialist.

Prior to attending the TMC, BN was seen by an otolaryngologist to rule out any underlying health-threatening condition requiring medical or surgical intervention. BN presented to the otolaryngologist with complaints of increased left-sided unilateral tinnitus over the past 2 weeks accompanied by left ear otalgia, which BN rated 5 on a 10-point pain scale (1 = no pain; 10 = worst imaginable pain). BN further reported that the ear pain increased when she used a cotton swab in her ear canal or the surrounding area—specifically the temporomandibular joint (TMJ) area. She also reported neck and upper back discomfort as well as previous episodes of vertigo. Physical examination and magnetic resonance imaging (MRI) findings were all within normal limits. Due to reported increased tinnitus and anxiety, the patient was prescribed 0.5 mg of alprazolam, to be taken by mouth at bedtime as needed for her tinnitus.

69.2 Questions to the Reader

1. In addition to ruling out or treating underlying health-threatening disease, what role should the physician play in the overall management process of patients with tinnitus?
2. Are drugs such as alprazolam effective in reducing tinnitus?

69.3 Discussion of Questions to the Reader

1. In addition to ruling out or treating underlying health-threatening disease, what role should the physician play in the overall management process of patients with tinnitus?
   By conducting a thorough case history and physical examination and ordering appropriate follow-up tests, physicians can help alleviate the patient’s fear that he/she might have a serious medical problem. This is a critical first step in the management process by facilitating acceptance and even habituation of the tinnitus. During the otolaryngologist’s workup and evaluation, however, it is important for the physician to avoid heightening the patient’s preoccupation with the tinnitus by creating unnecessary fear or anxiety about its underlying cause. This may, in fact, produce further anxiety about the tinnitus and exacerbate the patient’s perception of tinnitus and the subsequent emotional reaction to it. On the other hand, the physician should not minimize the presence of tinnitus or counsel the patient to “live with it!” Once any underlying health condition requiring intervention has been ruled out (or medically/surgically treated, if necessary), physicians need to provide encouragement to and reassure their patient that help may be available from other healthcare providers.
2. Are drugs such as alprazolam effective in reducing tinnitus?
   There are currently no medications approved by the U.S. Food and Drug Administration (FDA) specifically for treating tinnitus. A number of medications, however, are often prescribed by physicians to treat insomnia, anxiety, depression, phobias, obsessive-compulsive behaviors, or other psychological problems associated with the negative psychosocial consequences of tinnitus. Among the drugs prescribed, alprazolam has been shown to provide relief for tinnitus sufferers in a controlled double-blind study.

69.4 Audiological Testing

As part of the initial workup with the otolaryngologist, an audiological examination was conducted. Pure-tone audiometry revealed normal hearing sensitivity through 2000 Hz sloping to a mild high-frequency sensorineural hearing loss, bilaterally. Speech recognition thresholds (SRTs) were 15 dB HL bilaterally indicating normal ability to receive speech and were consistent with the pure-tone averages. Word recognition scores (WRSs) using Northwestern University Auditory Test No. 6 (NU-6) word lists in quiet revealed normal ability to recognize speech bilaterally. Tympanometry revealed normal pressure (daPa), ear canal volume (mL), and static compliance (mL) bilaterally. Ipsilateral and contralateral acoustic reflex thresholds (ARTs) were present at expected levels, and negative reflex decay was observed at 1000 Hz bilaterally. Consistent with BN’s history, increased ear pain/discomfort occurred when the 3A insert earphone was inserted or the TDH-50 P supra-aural headphone was placed on her ear to conduct audiometry as well as inserting the probe tip used for immittance measurements.

BN indicated she had been aware of the tinnitus for approximately 1 year; however, it has become more bothersome over the past few months without any apparent reason. When questioned about the nature of the tinnitus, BN reported her tinnitus typically consists of at least three sounds and has a “cricket-like” quality, although the tinnitus often suddenly changes to either a completely different sound or a different pitch. The changes can also involve the perception of loudness. Using a numerical rating scale, she indicated that pitch (1 = very low pitch; 10 = very high pitch) changed from a 7 to a 10 throughout the day. BN indicated a range of loudness on a Likert scale (1 = very faint; 10 = very loud), from a magnitude estimation of

3 to 10 over the course of the day. The perceived changes in tinnitus pitch and loudness were exacerbated by changes in head position, jaw movement (i.e., opening jaw increases loudness) and increased emotional or mental stress. It was also noted that when her tinnitus changed she also experienced an associated pain in and around her ear, rated 5 on a 10-point Likert pain scale (1 = no pain; 10 = worst imaginable pain). BN scored 78 out of 100 points on the Tinnitus Handicap Inventory (THI) classifying her in the severe/catastrophic tinnitus handicap category. The THI is a 25-item questionnaire quantifying the negative psychosocial consequences of tinnitus. For each item, the patient responds with a "yes" (4 points), "sometimes" (2 points), or "no" (0 points). Total scores range from 0 to 100 points with higher scores reflecting greater perceived tinnitus activity limitation and/or participation restriction.

69.5 Additional Questions to the Reader

1. In addition to providing information about the type and severity of hearing loss, audiometric configuration, and functional integrity of the auditory system, what important role does audiological examination serve in the tinnitus rehabilitation process?
2. What additional case history information is important to obtain during an intake interview with a patient with tinnitus?
3. What are the most unusual tinnitus complaints reported by this patient?

69.6 Discussion of Additional Questions to the Reader

1. In addition to providing information about the type and severity of hearing loss, audiometric configuration, and functional integrity of the auditory system, what important role does audiological examination serve in the tinnitus rehabilitation process?
   Audiological test results provide a basis for the selection of appropriate sound therapy. For example, a patient with hearing loss and tinnitus may benefit from fitting a combination unit (instrument that is a hearing device and a sound generator). In contrast, a patient with normal hearing sensitivity may benefit from the use of a sound generator alone. Using the THI and its companion form, the Hearing Handicap Inventory for the Elderly/Adults (HHIE/A), permits the audiologist (and patient) to assess which condition is more problematic, the hearing loss or the tinnitus. The HHIE/A is a 25-item questionnaire designed to evaluate the emotional and social/situational consequences of hearing loss using the same scoring response format as the THI. When used together, the THI and HHIE/A questionnaires help direct the clinician to the selection of devices to (1) compensate for the hearing loss, (2) provide relief for the tinnitus, or (3) provide management of both hearing loss and tinnitus.
2. What additional case history information is important to obtain during an intake interview with a patient with tinnitus?
   In general, the case history for patients with tinnitus should include the following:
   - Descriptive attributes of the tinnitus such as perceptual features (e.g., location, pitch, loudness), duration (e.g., onset, percentage of time disturbed or aware), and quality (e.g., ringing, hissing, roaring)
   - Noise exposure and drug use
   - Dental problems (e.g., jaw pain, teeth grinding)
   - Specific behavioral, social, emotional, and interpersonal consequences of tinnitus
   - Factors that may increase or reduce tinnitus disturbance
   - Perceived hearing, hearing loss, and issues with sound tolerance
   - Previous tinnitus treatment/s (e.g., medical/surgical/rehabilitative) and benefit from treatment/s
   - Prioritization of concerns (e.g., hearing loss, tinnitus, sound tolerance)
   - Expectations about treatment
3. What are the most unusual tinnitus complaints reported by this patient?
   Patients often report variability associated with the perceptual attributes of their tinnitus; however, BN reported that changes in pitch and loudness occurred with contraction of the head, neck, and/or jaw. Tinnitus that can be modified or modulated with changes in movement, pressure, or touch to the head and neck area is known as somatic tinnitus. Recall that BN reported the presence of left-sided ear pain with use of both audiometric transducers (super-aural headphones and insert earphones) and the immittance probe, but that the pain/discomfort was only in the left ear.

69.7 Additional Testing

As part of the TMC, BN underwent a multidisciplinary team assessment. Table 69.1 summarizes the findings following a brief examination (15–20 minutes) by each specialist on the team.

69.8 Additional Questions to the Reader

1. What types of stimulation produce somatic tinnitus?
2. What are myofascial trigger points (MTPs) and how do they relate to the patient's reported tinnitus and ear pain?
3. Are there anatomical connections between the auditory system and other sensory and motor systems?

69.9 Discussion of Additional Questions to the Reader

1. What types of stimulation produce somatic tinnitus?
   Somatic modulation of tinnitus has been achieved by a variety of different stimuli, including forceful muscle contractions of the head, neck, and limbs; eye movements in the horizontal and/or vertical plane; pressure on MTPs; cutaneous stimulation on the hand or fingertips; electrical stimulation of the median nerve; orofacial movements; and pressure

Table 69.1 Observations and findings following assessments by each specialist in the tinnitus management clinic

Specialty	Observations/findings
Audiology	• Immediate relief from tinnitus in the clinical setting using ◦ ear-level sound generators ◦ Neuromonics Oasis Device ◦ music played through MP3 player
Dentistry	• Loudness of tinnitus changed with teeth clenching, jaw thrust, and opening mouth • Loudness of tinnitus increased and reported pain in ear canal and temporomandibular joint (TMJ) area with palpation of masseter muscle (myofascial trigger point [MTP])
Neurology	• Left C1 (cervical spine) malrotation • Significant somatic modulation of tinnitus with manipulation of jaw and neck with the jaw manipulation producing greater increased tinnitus in comparison to neck manipulation
Physical therapy	• Increased tinnitus loudness with jaw movements and neck rotation • Overall, poor sitting and standing posture
Psychology	• Results of Patient Health Questionnaire-9 score (19 points; PHQ-9) suggested moderate to severe depression • Results of Generalized Anxiety Disorder-7 score (11 points; GAD-7) suggested moderate anxiety

applied to the TMJ. Patients may also report temporary changes in tinnitus occurring during common daily movements of the jaw or neck (e.g., clenching teeth as in the case of BN) or by applying pressure on the temples, mandible, cheek, mastoid, or neck with their fingertips. During the TMC screenings, physical examination by the dentist, physical therapist, and neurologist included an active search for tinnitus modulation through different stimuli, including digital pressure of the MTPs and movements of the neck, shoulder, mandible, and eyes (to detect gaze-evoked tinnitus).

2. What are MTPs and how do they relate to the patient's reported tinnitus and ear pain?
   MTPs are hypersensitive spots within bands of skeletal muscle fibers that can cause local or referred pain with well-defined patterns for specific muscle groups. MTPs are considered "active" when their stimulation causes an increased referred pain pattern that is similar to the patient's pre-existent pain complaint. As in the case of BN, trigger points underlying the deep layer of the masseter muscle resulted in both tinnitus and referred pain to the ipsilateral ear during muscle palpation, contraction, and jaw maneuvers. Interestingly, masseter muscles are among the first muscles to show a physical response to emotional stress. That is, in response to stress, the masseter muscle contracts and remains contracted for long periods of time. In this connection, BN reported emotional distress, which was supported by her moderate to severe scores on the Patient Health Questionnaire-9 (PHQ-9) and Generalized Anxiety Disorder-7 (GAD-7) (□ Table 69.1). Accordingly, hyperactivity of the masseter muscle, especially during high levels of emotional anxiety and distress, may exacerbate both the tinnitus and the pain.
3. Are there anatomical connections between the auditory system and other sensory and motor systems?
   Somatic tinnitus appears to be a good example of central integration of the central nervous system; that is, an auditory perception is modulated through nonauditory stimulation/manipulation. For example, abnormal somatosensory feedback from muscle spindles or golgi tendon muscles in the head and neck muscles may release the dorsal cochlear nucleus from inhibition, resulting in tinnitus. In animal studies, trigeminal and dorsal root ganglia have been shown to relay afferent somatosensory information from the peripheral to auditory centers in the brainstem, which both excite and inhibit cochlear nucleus units. The modulation of firing rate and synchrony in the dorsal cochlear nucleus by such somatosensory input has been correlated with somatic tinnitus.

69.10 Diagnosis and Recommended Treatment

Physical examination of BN by the dentist, neurologist, and physical therapist was consistent with somatic tinnitus. That is, BN's tinnitus could be modulated during masticatory muscle contraction, jaw clenching, and manipulation of the neck and cervical spine. In addition, both left-sided tinnitus and otalgia were increased by stimulation of MTPs during compression of the masseter muscle.

Based on results of the multidisciplinary team assessment, several recommendations were provided to BN:

1. A dental orthotic (i.e., a bite splint) was recommended to improve jaw mechanics, including reduction of jaw contraction to decrease hypercontraction of the masseter muscle.
2. Self-help therapies were also provided to help decrease clenching and overloading of the masticatory system to promote MTP deactivation. For example, BN was instructed to avoid gum chewing, cheek biting, nail biting, and eating "hard chewy" foods. In addition, she was counseled about proper tongue and teeth posture (i.e., lips together, teeth apart, tongue resting lightly on the palate).
3. Physical therapy was recommended to address MTP release techniques (e.g., self-massaging of the MTPs) and posture retraining.
4. A relaxation CD, developed by the Department of Psychiatry and Psychology at the Cleveland Clinic, was provided to promote progressive muscle relaxation and reduce muscular tension. This CD contains several tracks of guided relaxation exercises with soothing background music. There are a number of similar commercially available CDs that can be purchased via the Internet or in retail stores. Further, a workbook focusing on acceptance and commitment therapy (ACT)

was recommended, titled The Mindfulness & Acceptance Workbook for Anxiety. This self-help workbook offers guidance in anxiety management (a major underlying response to tinnitus), helping people to accept the difficulties that come with life. Reduction of anxiety through acceptance helps to reduce the symptoms of somatic tinnitus by further promoting relaxation, thereby reducing hyperactivity of the head, neck, and jaw muscles.

5. The benefits of sound therapy (e.g., personal sound generators, Neuromonics Tinnitus Treatment (Neuromonics, Westminster, CO), use of applications with personal music player, tablet, or Smartphone) were discussed. Selection of a specific form of sound therapy is based on a number of patient-focused factors, including lifestyle, manual dexterity, acceptability of the auditory stimulus, cosmetics, daily dosage (hours/day of sound therapy usage recommended), and personal finances. For BN, use of sound therapy would serve as a palliative treatment by producing tinnitus relief.

69.11 Outcome

After the initial visit at TMC, BN scheduled follow-up appointments with the dentist and physical therapist for comprehensive examinations. The coordinated treatment plans developed by those providers focused on deactivation of masticatory muscle MTPs; self-help therapy to recognize and discontinue daytime teeth/jaw clenching; and physical therapy to address decreased range of motion in the cervical and thoracic spines, and tenderness of cervical and jaw muscles. Use of the dental orthotic and ongoing physical therapy essentially resulted in a "cure" for BN. Following 7 months of treatment BN indicated she was "no longer bothered by the tinnitus" and did not experience otalgia in her left ear. Readministration of the THI resulted in a score of 12/100 points, far exceeding the 20-point change from the initial THI score (78/100 points) considered to be a statistically and clinically significant reduction in perceived tinnitus activity limitation/participation restriction.

69.12 Key Points

1. Evidence is accumulating that the perception of tinnitus is multimodal, arising from complex interactions among different sensory and motor systems.
2. The hallmark of somatic tinnitus is a temporary change in the psychoacoustic characteristics (e.g., pitch, loudness, quality) of tinnitus modulated by some sort of stimuli, typically involving manipulation or contraction of head, neck, and/or jaw muscles.
3. A subgroup of patients with somatic tinnitus exists requiring assessment and intervention by a multidisciplinary team including audiology, dentistry, neurology, physical therapy, and psychology.
4. It is critical that audiologists establish ongoing professional relationships with the aforementioned providers thereby viewing the management of tinnitus from a broader and more comprehensive perspective.

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Part 8

Other Rehabilitative Strategies

70 It’s Not All About the Hearing Aid 310

71 Exostoses, Hearing Aids, and Brain Training 315

70 It's Not All About the Hearing Aid

Jennifer P. Taylor and Lisa Lucks Mendel

This case addresses issues that a clinician should consider when the audiological rehabilitation needs of the patient extend well beyond the hearing loss and hearing aids.

70.1 Clinical History and Description

JL is a 76-year-old male who scheduled an appointment for a follow-up audiological examination due to continued hearing difficulties in a variety of listening situations. He has worn bilateral hearing aids for 15 years, and it has been 2 years since his last audiological examination. JL was accompanied by his wife and adult son who recently returned to live with his parents. JL wears a behind-the-ear (BTE) digital hearing aid in his right ear that he purchased 7 years ago and a BTE in his left ear that was purchased 12 years ago. JL's wife is interested in pursuing new hearing aids to help JL hear better and to allow her and JL to maintain a comfortable volume on the television. JL's wife reports that "JL's hearing difficulties are the hardest thing they have had to deal with in their 53 years of marriage." JL's son reports he is concerned that his relationship with his father is deteriorating because his father's inability to hear well with his current hearing aids has caused numerous misunderstandings and arguments within the family. JL reports he does not feel new hearing aids will help because he believes his wife and son "tell secrets so that he cannot hear what they are saying." JL's wife reports she and her son were not telling secrets, but were discussing matters they did not want their 9-year-old granddaughter to hear. Ironically, JL reports that his 9-year-old granddaughter is the only person who communicates with him properly because "everyone else talks too fast, does not speak clearly, and mumbles." JL has a history of noise exposure from his years as a farmer. He also had a melanoma on the helix of his right pinna that was removed. JL believes the earmold he was using may have caused the melanoma.

70.2 Audiological Testing

Results from an audiological examination revealed a moderately severe to profound sensorineural hearing loss in the right ear and a moderate to profound sensorineural hearing loss in the left ear (Fig. 70.1). Speech recognition thresholds (SRTs) were 75 and 65 dB HL in the right and left ears, respectively, indicating a severe loss in the ability to receive speech in the right ear and a moderately severe loss in the ability to receive speech in the left ear. Word recognition scores (WRSs) presented at 20 dB sensation level (SL) re: SRT were obtained using full Central Institute for the Deaf (CID) W-22 word lists recorded by a male talker. Word recognition lists were presented at reduced SLs due to patient report of loudness recruitment. WRSs reflect very poor ability to recognize speech in a quiet environment bilaterally. The WRSs revealed a significant decrease in word recognition from the previous audiological evaluation completed 2 years ago. SRTs were in agreement with pure-tone averages suggesting good intratest reliability. Tests of word recognition in noise were not attempted due to the patient's difficulty and frustration during word recognition testing in quiet. Immittance audiometry revealed Jerger type A tympanograms with normal ear canal volume (0.6 mL right ear; 0.5 mL left ear), static admittance (1.4 mL right ear; 1.6 mL left ear), and tympanometric peak pressure (10 daPa right ear; − 10 daPa left ear). The results suggest normal middle ear function, bilaterally. Acoustic reflex thresholds (ARTs) were absent bilaterally to contralateral and ipsilateral stimulation due to the magnitude of the hearing loss.

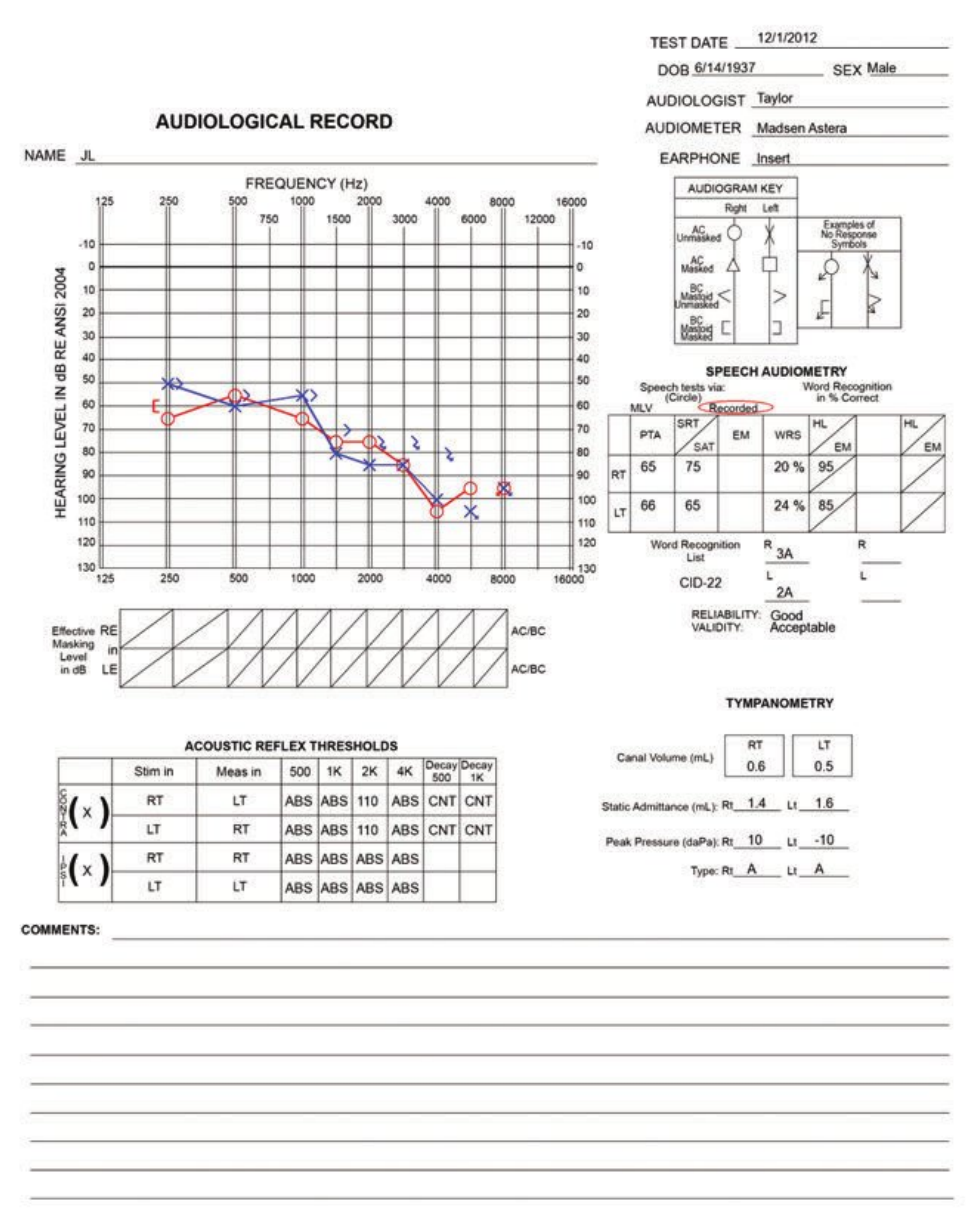
Throughout the audiological examination, JL displayed considerable anger toward the clinician and complained about having to complete so many tests. After word recognition testing was completed, JL stated, "I wanted to quit the test because it was a waste of time since I can't hear anything." Before the clinician could begin counseling JL and his family regarding his test results, he reported the reason he had so much trouble hearing was because everyone else did not communicate well with him. Despite JL's refusal to discuss any new options for amplification, JL's wife and son persisted in learning what options were available to improve his hearing because of the difficulties the family was experiencing as a result of JL's hearing loss.

70.3 Questions to the Reader

1. What information is present in the case history and test results that will impact a clinician's decision making for JL?
2. What recommendations can a clinician make regarding amplification options for JL?
3. What recommendations can a clinician make regarding audiological rehabilitation for JL and his family?

70.4 Discussion of Questions to the Reader

1. What information is present in the case history and test results that will impact a clinician's decision making for JL?
   Several questions need to be asked and then answered regarding the concerns raised during the taking of the case history to determine how this information may relate to the results of the audiological examination. With regard to JL's history of noise exposure with loud farming equipment, did JL use hearing protection during his years as a farmer? What type(s) of hearing protection (disposable or custom earplugs and/or muffs) were used? Is he currently exposed to loud noises and is he using hearing protection? The clinician should also consider if JL's noise exposure is a contributing factor to his decreased speech recognition abilities or whether there could be other reasons for the decline. The clinician should also review previous audiological examination



AUDIOLOGICAL RECORD

NAME JL

TEST DATE 12/1/2012

DOB 6/14/1937 SEX Male

AUDIOLOGIST Taylor

AUDIOMETER Madsen Astera

EARPHONE Insert

SPEECH AUDIOMETRY

Speech tests via: (Circle) MLV [✓ Recorded]

Word Recognition in % Correct

	PTA	SRT / SAT	EM	WRS	HL / EM		HL / EM
RT	65	75		20 %	95		
LT	66	65		24 %	85		

Word Recognition List R 3A L 2A R L

CID-22

RELIABILITY: Good

VALIDITY: Acceptable

TYMPANOMETRY

	RT	LT
Canal Volume (mL)	0.6	0.5

Static Admittance (mL): Rt 1.4 Lt 1.6

Peak Pressure (daPa): Rt 10 Lt -10

Type: Rt A Lt A

ACOUSTIC REFLEX THRESHOLDS

	Stim in	Meas in	500	1K	2K	4K	Decay 500	Decay 1K
CONTRA (x)	RT	LT	ABS	ABS	110	ABS	CNT	CNT
	LT	RT	ABS	ABS	110	ABS	CNT	CNT
IPSI (x)	RT	RT	ABS	ABS	ABS	ABS		
	LT	LT	ABS	ABS	ABS	ABS		

COMMENTS:

Fig. 70.1 Audiometric findings.

results and determine the long-standing nature of the hearing loss. Has there been a continual decrease in hearing thresholds and WRSs? Is the amount of progression greater than would be expected? The clinician must also investigate why the continued decrease in WRS is affecting JL’s attitude as much as it is.

Given the history of melanoma, the clinician should determine if JL is being followed by physicians for any other health-related issues. Also, did JL receive any chemotherapy and/or radiation therapy, which may help explain the decrease in hearing? It is possible that JL’s emotional response to his hearing loss could be exacerbated by such health-related issues leading to conditions such as poor general health, reduced interpersonal communication, and depression. In addition to health concerns, the clinician should explore whether JL’s anger stems from home-related issues such as his son and granddaughter moving in with him and his wife. If the clinician determines that these factors may be playing a role in JL’s anger and frustration, then how can these health-related factors best be addressed? Does the clinician need to consider referring JL to a psychologist for further evaluation and counseling?

The clinician should also probe further about JL’s motivation for scheduling the appointment. Was it to appease his wife and son? If so, the clinician must consider the fact that JL’s anger and lack of motivation may not be conducive to a successful fitting with new hearing aids. The clinician should also determine what activity limitations and participation restrictions JL has due to his hearing loss and decreased speech recognition abilities.

The clinician must consider the comments made by JL’s family during the case history regarding their communication difficulties. The statements made by JL’s wife indicate the communication between her husband and herself has been seriously impacted by JL’s hearing loss. JL’s son has also experienced a similar negative impact in his relationship with his father. The role of the family in seeking solutions to JL’s hearing difficulties cannot be underestimated nor discounted here. The many concerns the family raised in the case history must be addressed by the clinician when determining an intervention plan for JL. Although the hearing loss may be the underlying cause of the problem, the poor communication, anger, and frustration on the part of the entire family must be the focus of the intervention plan. JL’s comments in the case history reflect a considerable level of distrust within his family unit. The emotional issues that JL is experiencing are likely the direct contributors toward his suspicions, anger, lack of motivation, sadness, and denial. Unproductive learned behaviors such as maladaptive passive communicative behavior on the part of the family and aggressive behavior on the part of JL make the situation worse.

2. What recommendations can a clinician make regarding amplification options for JL?

   JL's poor speech recognition scores suggest the possibility that cochlear implants could be an option. However, due to some specific health concerns, JL decided not to participate in an evaluation for candidacy for cochlear implants.

   Before determining if newer hearing aids will benefit JL, the clinician needs to determine if his current hearing aids are meeting manufacturer specifications and providing appropriate amplification via real ear measures. The clinician should also consider JL's success with his current hearing aids because JL is a previous hearing aid user. If the clinician believes that new hearing aids could be beneficial, would JL benefit from adaptive and automatic features in the hearing aids such as directional microphone technology? Adaptive and automatic directionality would allow JL the freedom to participate in dinner conversation without having to focus on making adjustments to his hearing aids.

   Frequency-lowering technology may also provide benefit to JL given the continued decrease in his high-frequency hearing thresholds and may improve his speech recognition abilities. Perhaps these advanced features could be incorporated into a digital power BTE to provide the necessary gain JL requires. A remote control could be considered to allow JL to maintain some control over his listening situations. Administration of a hearing aid benefit self-report tool, such as the Client-Oriented Scale of Improvement (COSI), would be extremely beneficial for JL to complete along with his family to document specific situations where they would like to see improvement. Lastly, hearing assistance technology (HAT) may be considered in addition to new hearing aids given JL's wife's comment about the volume of the television. The clinician should pursue other aspects of JL's lifestyle to determine if other HAT would also be helpful to provide benefit in his everyday activities and communication.

3. What recommendations can a clinician make regarding audiological rehabilitation for JL and his family?

   The clinician needs to develop a clear plan for intervention. To do this, the clinician must consider all the communication comments and observations made throughout the case history, audiological examination, and counseling. The comments made by all members of the family indicated very poor communication with JL, and as a result, JL and his family have learned to use poor communication behaviors. In addition, the clinician should evaluate JL's responses to the COSI to help guide the rehabilitation program and determine what other treatment options might be beneficial for JL. If additional information is needed about JL's activity limitations and participation restrictions, a number of other self-assessment questionnaires are available. These include the Self-Assessment of Communication (SAC) and the Hearing Handicap Inventory for the Elderly (HHIE). The clinician should also consider assessing the family's views of JL's limitations with communication by using questionnaires designed for significant others, such as the Significant Other Assessment of Communication (SOAC).

   All of the evaluation measures, questionnaires, and discussions with JL and his family suggest to the clinician that audiological rehabilitation is critical for JL to achieve optimal success in communication. Further, the clinician should strongly recommend that all members of the family participate in an audiological rehabilitation program. The clinician should consider group versus individual therapy options to determine which would be most beneficial for JL and his family. If group therapy is selected, the clinician needs to consider if the family will be able to openly discuss their communication difficulties. Will hearing other families discuss their communication difficulties make it easier for JL and his family to realize that their problems are not unique to them? Will the group setting allow JL, his wife, and son the ability to effectively discuss and express their emotions about JL's hearing loss and its impact on their family dynamics? Perhaps such openness might allow other families in the group sessions to have a greater appreciation for their own communication challenges as well. Individual therapy may also be warranted with a focus on more specific strategies for JL and his family to practice and incorporate into their daily lives.

   Once the clinician decides if group or individual audiological rehabilitation will be recommended, then the clinician must consider what the focus areas will be for each aspect of rehabilitation. The clinician should develop a program consisting of many essential behavior modification components, including communication breakdowns, facilitative and repair strategies, relaxation strategies, and speechreading exercises. Initially the clinician will need to facilitate a discussion with JL and his family regarding their past communication difficulties. Further, the clinician should encourage the family to discuss the emotions experienced when those communication difficulties occur. JL needs to learn to take ownership of his hearing loss and accept the important role it has in his communication. JL's wife and son also need to recognize the roles they have played in the poor communication they have had to date. Recognizing the causes of the communication difficulties and negative reactions that result from them will help JL and his family be informed regarding how to address future communication difficulties in a more positive and productive manner. The family also needs to agree to forgive past behaviors and create a plan for moving forward.

   As the family learns how to deal with communication difficulties, the clinician should also introduce various facilitative and repair strategies to help the members of the family take greater control of the communication situation. For example, in a noisy situation, the family needs to learn to move to a quieter area before initiating communication with JL. Making JL and the family more aware of their surroundings allows the members of the family to control their environment, which may result in more successful communication opportunities. In addition, because JL has many complaints regarding communicating with others, managing his anger, and feelings of exclusion, these concerns must be addressed to help JL better accept his hearing loss. The clinician must counsel JL and his family on how to recognize anxious behaviors during communication and learn to implement appropriate relaxation techniques in such situations.

Due to the nature and extent of JL's hearing loss and communication problems, the clinician should consider speechreading exercises as part of the intervention plan. Improvement in his speechreading ability may give JL more confidence in communication and should improve the amount of information he receives. Having his family participate in these exercises is a critical factor in order for the family members to appreciate the challenges that JL faces daily. Further communication training (e.g., auditory/perceptual training such as the Listening and Communication Enhancement (LACE) program may also be beneficial for improvement in JL's performance.

70.5 Diagnosis and Recommended Treatment

JL has a moderate to severe sloping to profound bilateral sensorineural hearing loss with very poor word recognition abilities. Information obtained during the case history and audiological examination from JL and his family clearly suggest that his anger and frustration about his hearing loss are a hindrance to his and his family's ability to move forward. The clinician's focus must first be on a complete intervention program that emphasizes counseling and rehabilitation strategies in addition to exploring new amplification options. The clinician did recommend JL be seen for a hearing aid evaluation; however, it was critical to begin audiological rehabilitation as soon as possible due to the negative interactions occurring within the family and their maladaptive communication behaviors. The clinician discussed various options for individual and group family therapy and using the LACE program to guide JL to promote listening strategies. The clinician then administered the HHIE and COSI to assist in developing a detailed plan for intervention. When the clinician made these recommendations, JL's wife and son were in agreement with this strategy; however, JL was less than enthusiastic.

70.6 Outcome

The family made the decision, after considering all of their options, to enroll in a 6-week group audiological rehabilitation program, which included speechreading and communication strategy training. Other topics included in the group sessions focused on understanding the audiogram, relaxation techniques, and communication problems. Each family expressed their particular concerns about communication, and initially JL's participation was limited. He overtly expressed anger and frustration during the first session. Efforts were made by other group members to engage him in the conversation and speechreading exercises; however, JL was still resistant. During the first speechreading session, families were asked to share their reasons for attending the session and what they hoped to gain from the experience. JL reported his only goal was that his son and wife would learn to better communicate with him. He was unable to see his role in the process. In contrast, the other group participants expressed the understanding that the communication difficulties were a shared problem and that all parties would need to learn strategies that they could use to improve communication. The positive attitude demonstrated by the other group members began to have an impact on JL. Even though it was hard for him to change his longtime behaviors, JL started to evaluate the importance of his role in successful communication.

During the first speechreading exercise, which was very difficult for him, JL became angry and did not want to participate. Over the next couple of weeks in the program, the other participants and facilitator noticed that JL became more excited during the sessions as he began to demonstrate improvement in his speechreading ability. It was clear that the group dynamic allowed JL to experience more positive communicative behaviors. Much of this was facilitated by the fact that the other group participants took an active role in "counseling" JL and his family. Eventually, JL started to compete against his wife and son and other members of the group during the activities and began having fun with the entire experience. By the final speechreading activity JL's performance improved from 0 out of 10 to 9 out of 10 correct. JL gained confidence with his communication and began implementing the communication strategies outside the group session, which allowed him to communicate more freely with his family. The impact on the family was significant, and during the final session JL's wife stated "this was the best thing to happen since he was diagnosed with hearing loss."

JL is currently scheduled for a hearing aid evaluation to determine if current technology would provide greater benefit than his current hearing aids in his daily communication. This will allow the clinician to discuss the various options for more current hearing aids. It was also recommended that JL apply for an amplified telephone provided through the state communication authority program and consider wireless accessories for use in the car, with the television, in restaurants, and in other listening situations.

This case demonstrated how the clinician and JL's family united to develop a plan to help JL learn to cope with his hearing loss, and at the same time, provide JL an outlet to openly discuss his problems. The group dynamic of the rehabilitation program allowed JL and his family to feel nurtured and safe while discussing their communication issues. As a result, anger no longer ruled JL's communication, and his family dynamic improved significantly. In this case, JL's rehabilitation focused on addressing the emotional issues hindering his progress showing that his success was not completely dependent on amplification. A shift in his attitude and the elimination of his maladaptive behaviors allowed JL to take more responsibility for his communication and improved his family dynamic.

70.7 Key Points

1. Clinicians must listen carefully during the entire diagnostic appointment for opportunities to counsel patients regarding emotional issues that may hinder progress.
2. Sometimes clinicians must address the emotional issues affecting the patient with hearing loss before considering amplification.
3. Emotional reactions to hearing loss need to be addressed throughout the evaluation and rehabilitation process.

4. Family and support groups are key factors in successful rehabilitation programs.
5. Hearing loss affects the entire family.

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71 Exostoses, Hearing Aids, and Brain Training

Therese C. Walden

This case report discusses the importance of periodic audiological examination in the presence of stable hearing loss, decreasing signal to noise ratio (SNR) ability, and age-related cognitive decline.

71.1 Clinical History and Description

EW is an 86-year-old male who presented in the clinic more than a decade ago for a diagnostic audiological examination. The patient had a long history of noise exposure while serving in the military as a Navy pilot. His medical history was negative for familial adult-onset hearing loss or any medical issue that could contribute to the hearing loss. There was no history of medications (prescribed or over-the-counter) that may have contributed to the hearing loss. At 74 years of age, the patient reported slight hearing difficulties in daily listening situations: hearing his wife, some social situations, and listening to the television. He also reported being an avid swimmer in his youth and upon otoscopic examination exostoses were identified bilaterally in his ear canals. The exostoses in the right ear were greater in size and number than noted in the left ear. Exostoses are a reactive tissue process borne of long-term exposure to cold water or cold air. They are excessive growth of the tympanic bone; typically seen in multiples, in both ear canals and are benign. Patients are generally asymptomatic unless the growths become large enough to block the ear canal and cause hearing loss or recurrent infection. In addition to the identification of the exostoses in the ear canals, EW was diagnosed with bilateral mild to moderate hearing loss and amplification options were discussed. Initially, EW chose not to treat the hearing loss, and there was no recommended treatment for the exostoses. Periodic audiological testing, approximately every 2 years, indicated a fairly stable hearing loss (air conduction thresholds) in each ear. The patient eventually tried one custom-made hearing aid, which he wore for nearly 6 years with reported success. Eventually speech-in-noise testing was added to the periodic evaluations. Although the patient performed within normal limits initially, his speech-in-noise ability began to deteriorate. Binaural hearing aids were eventually used, and the patient began at-home brain-training exercises to help, primarily, with his memory skills. The complementary effects of consistent binaural hearing aid usage and cognitive training resulted in a positive outcome for the patient in terms of improved auditory function and processing.

71.2 Audiological Testing

At the initial evaluation in 2000, EW was diagnosed with bilaterally symmetrical mild to moderate sensorineural hearing loss. Immittance audiometry indicated normal tympanograms and normal acoustic reflex thresholds (ARTs) through 2000 Hz. Word recognition testing (in quiet) at a comfortably loud level was within normal limits. After careful counseling on the benefits of amplification as a treatment option for his hearing loss, the patient deferred hearing aid use in 2000. No treatment was recommended for the bilateral exostoses. At his next appointment in 2002 (the patient had not returned in 2001), the hearing loss was found to be stable and the potential effects of long-term untreated hearing loss (reduced ability to recognize sounds, reduced processing speed, etc.) were discussed with the patient. A completely in-the-canal (CIC) hearing aid was prescribed for the left ear. (The patient's preference was for a custom-fit hearing aid that was 'less noticeable'.) The size of the growths in the right ear prevented the adequate use of amplification in that ear. The left aid required minor modifications (use of a rotary tool and a sanding bit) to reduce the overall circumference of the aid to fit around the exostoses in the left ear canal. In 2004 and 2006, adding a hearing aid for the right ear was recommended, but the patient declined. He was, by self-report, successful with the monaural left hearing aid as the aid helped in listening situations where there was soft speech and for listening to the television. The patient also reported that the physical fit and comfort of the hearing aid (with the exostoses) was good. Additionally, at the 2006 examination, the audiologist performed speech-in-noise testing using the QuickSIN (Etymotic Research, Inc., Elk Grove Village, IL). The QuickSIN provides a clinical estimate of the patient's ability to hear speech (sentences) in the presence of background noise (four-talker babble). Six sentences with five key words in each sentence are presented at preset signal to noise ratios, which range from easy (+25 dB SNR) to difficult (0 dB SNR). The more keywords repeated back correctly, the lower the score. A score of approximately 2 dB SNR (or lower) is considered within normal limits. The patient's performance was within the normal range (0.5 dB SNR at 65 dB HL) under earphones with bilateral input.

In 2009, EW's hearing was once again examined, and his hearing had remained stable for nearly a decade. Additionally, there was no change in his word recognition scores in quiet (remained within normal limits) and the tympanograms also remained normal. He reported he had been wearing the left monaural CIC hearing aid intermittently with decreasing benefit. His QuickSIN score decreased from normal (0.5 dB SNR) to a mild to moderate SNR loss (5.5 dB at 65 dB HL). This increase in SNR loss was likely due to long-term "undertreated" hearing loss (i.e., monaural amplification) and the effects of aging on his auditory processing abilities. Upon otoscopic inspection, the exostoses appeared to have decreased in size bilaterally. With regard to hearing aid technology, miniature behind-the-ear (BTE) aids with domes were available for fitting. Additionally, EW reported that he had started an online brain training program called Brain Fitness—recently renamed BrainHQ (Posit Science Corp., San Francisco, CA: http://www.positscience.com) in 2008. The patient was interested in "training his brain" to improve memory, focus, and processing speed. These are skills that are negatively impacted not only by age-related cognitive decline but also by the effect of long-term hearing loss. The online brain training program consists of modules that "exercise" the brain in five areas: auditory sequencing (Sound

Replay), auditory memory (Match It), working memory (Listen and Do), long-term memory (Story Teller), and sound discrimination (Tell Us Apart). The patient was able to access an online report of his progress after completing 40 sessions, and the report was uploaded to his electronic medical record (EMR) by his audiologist.

Bilateral open-fit mini-BTEs with medium nonoccluding domes were fit. The domes easily slid into the ear canals, past the exostoses. Comfort and retention were good. The patient also noted a substantial subjective improvement in his overall hearing ability in most daily listening situations with the bilateral hearing aids. EW averaged 10 hours of use per day (based on subjective report and datalogging from the hearing aids). In 2011 and late 2012, speech-in-noise testing (QuickSIN) indicated a fairly stable SNR loss of 4.5 dB at an average conversational level of 55 dB HL. EW had continued use of the online brain training program, completing approximately 40 sessions per year. Table 71.1. summarizes the progress EW has made from 2008 to 2012 in percent improvement from baseline (2008) and then from session to session in the five modules. The Posit Science Corporation auditory program claims, based on published studies, to increase an individual's ability to absorb more information, which will improve auditory processing and lead to improved interpersonal interactions during daily communication. The effect of the aging process on peripheral hearing as well as the effect of aging on cognitive capabilities is well documented (see Suggested Reading section). Mild to moderately severe sensorineural hearing loss, for the most part, has no effective medical or surgical treatment but is amenable to audiological treatment through hearing aids and assistive devices (e.g., amplified telephones, alerting devices, personal FM systems, wireless remote microphones, and others). Audiological treatment of hearing loss, especially if initiated early, can reduce the adverse effects of hearing loss that can negatively affect communication ability, quality of life, and cognitive function, especially in the older adult. Higher-level cognitive processing is dependent on effective peripheral input so that the higher-level hearing functions of listening, comprehending, and communicating are not compromised by inconsistent auditory input. The effects of aging can also impact the peripheral auditory system, the higher-level central auditory system, and overall cognitive functions.

Table 71.1 Percent improvement on test scores from 2008 to 2012

2008 (Baseline)	2009	2011	2012	Brain training task
36%	1%	No change	No change	High or Low: Processing speed
41%	8%	No change	14%	Tell Us Apart: Sound discrimination
15%	3%	18%	4%	Match It: Auditory memory
19%	13%	No change	No change	Sound Replay: Auditory sequencing
9%	1%	No change	No change	Listen and Do: Working memory
35%	25%	24%	24%	Story Teller: Long-term memory

Note: Each year represents 40 sessions over approximately 9 months and 15 minutes/online work per day. Data from 2010 not available.

71.3 Questions to the Reader

1. What benign growths other than exostoses can be found in the ear canal?
2. What are the effects of peripheral hearing loss on the accuracy of auditory processing, especially in the older patient?

71.4 Discussion of Questions to the Reader

1. What benign growths other than exostoses can be found in the ear canal?
   Osteoma (pl. osteomata) is a differential diagnosis for exostoses. Osteomata are benign, singular bone lesions that can be spongy or more firm to the touch. The outer layer is made of a dense squamous epithelium with an underlying periosteum. The inner structure is composed of distinct fibrovascular channels surrounded by flat bone. Patients are generally asymptomatic unless the growth becomes large enough to block the ear canal and cause hearing loss or recurrent infection.
2. What are the effects of peripheral hearing loss on the accuracy of auditory processing, especially in the older patient?
   The accuracy of processing auditory signals is distorted due to peripheral loss. There is a reduced ability to distinguish speech from noise, reduced effective auditory memory (difficulty keeping up with the conversation), and reduced localization ability or timing, which affects processing speed.

71.5 Outcome

At his most recent visit, EW was fit with new open-fit mini-BTE hearing aids that were paired wirelessly with a remote microphone to further enhance EW's ability to hear speech in a background of noise (such as hearing his wife at home or at a restaurant) and improve listening to the television with the remote microphone placed near the loudspeaker of the television. The patient reports he is performing better on the brain training exercises overall, and he is hearing subtleties in speech and conversations that he did not hear previously. This improvement is perhaps due to consistent use of his bilateral hearing aids and the time spent on brain training. Currently, there is no method available to determine the contribution of hearing aid use and brain training; however, the benefit of both is clear for this patient. The use of mental skills exercises as a strategy to improve cognitive abilities has increased in the last few years, with many online programs available for use with personal computers, tablets, smart phones, and electronic readers. In addition to auditory training, there are training exercises that claim to improve intelligence, visual processing ability (correlates with the auditory training), gaming, and much more. Many programs are adaptive and adjust with the individual's progress in terms of difficulty. The use of brain training has replaced traditional memorization to improve memory and instead focuses on functional improvements of higher levels of cognitive processing. Brain training appears to serve as a complement to the effective use of amplification to help diminish cognitive decline associated with long-term hearing loss and aging.

71.6 Key Points

1. Consistent, annual (or biennial) audiological examinations are critical because they allow patients to understand how their hearing, auditory processing, and cognitive abilities are related and how these may change over time.
2. The use of amplification (and other contemporary devices) to treat hearing loss, especially early treatment, may help diminish the effects of aging (memory loss, inability to focus, decreased processing speed) oftentimes experienced by older adults.
3. Adjusting to patients' changing needs as they age and even challenging patients to try new treatment options is an important tool in the audiologist's arsenal, especially when these treatment options are supported by the evidence.

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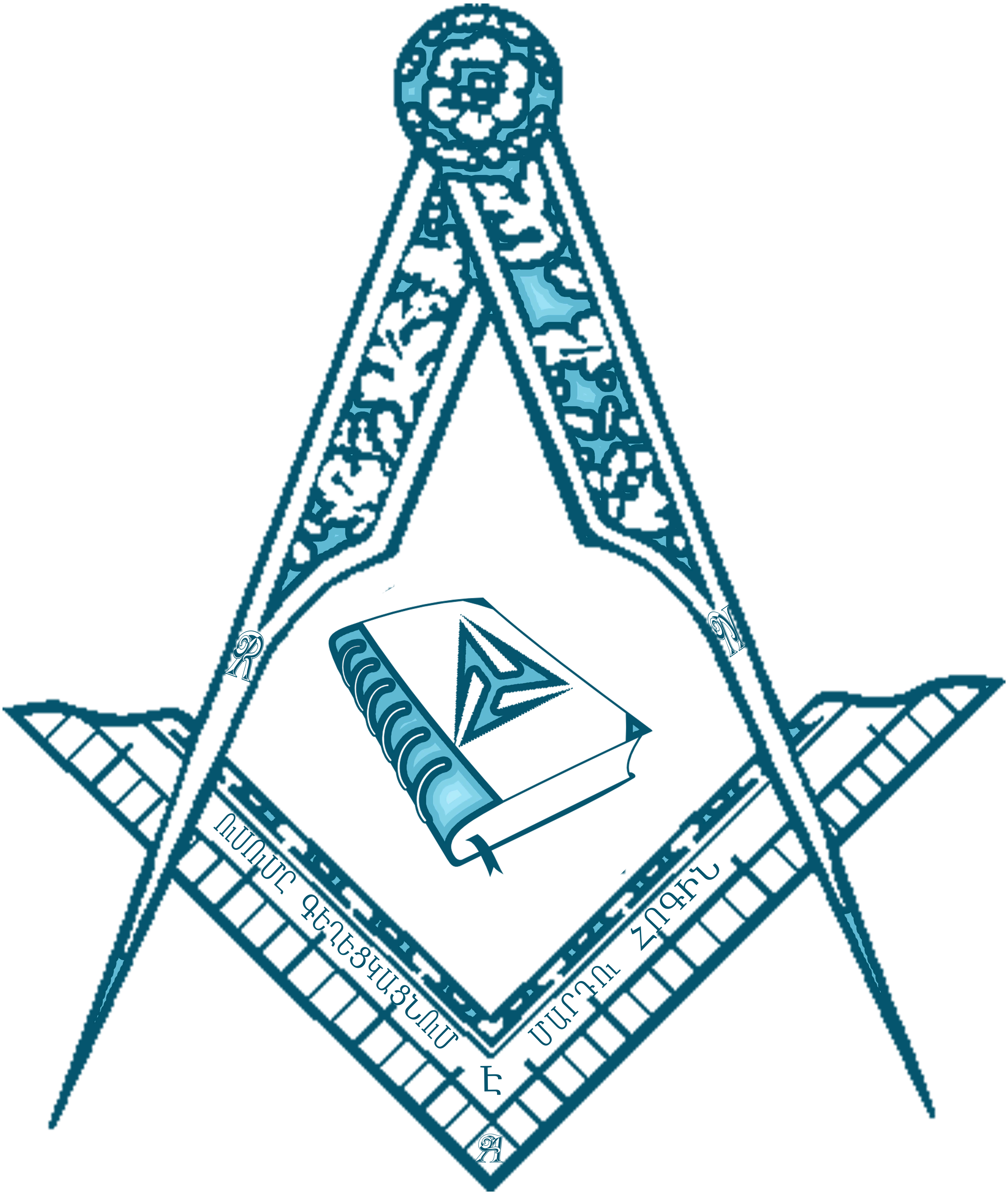
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